CALIFORNIA CANCER COMMISSION

SEMIANUAL SLIDE CONFERENCE

ON

NEOPLASMS OF THE EYE AND ADNEXA

MODERATOR:

A. R. IRVINE, JR., M.D., CO-DIRECTOR

ESTELLE DOHENY EYE FOUNDATION
LOS ANGELES, CALIFORNIA.

CHAIRMAN:

George J. Hummer, M.D.,
Santa Monica, California.

SUNDAY, December 8th, 1957.

9:00 A.M. - 4:00 P.M.

HOTEL UTAH
South Temple and Main Streets,
Salt Lake City, Utah.

Please send in your diagnoses, using the separate sheets enclosed, on or before November 25th, 1957, so that they may be tabulated before the meeting.

Please bring your protocol, but do not bring slides or microscopes to the meeting.
PLEASE NOTE:

There are multiple histories on Case Nos. 6, 12, 14, 18 and 19. Due to insufficient material on these cases, it was necessary to prepare sections from two to four different cases presenting the same diagnosis. For instance, you may have a slide that is numbered 60 in your set. This will be your Case No. 6 slide.
CONTRIBUTOR: A. R. Irvine, Jr., M.D., Estelle Doheny Eye Foundation, 272 South Lake Street, Los Angeles 57, California.

CASE NO. 1
December 8, 1957.

ACCESSION NO. 9417
OUTSIDE NO. EDEF 316-49

NAME: C. L.
AGE: 32 SEX: Female RACE: Cauc.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: In July, 1949, the patient noticed a blurring of the temporal vision in the right eye. The visual acuity was normal. A large, gray-brown, solid appearing detachment of the retina was seen in the posterior fundus on the nasal side of the optic disc. A clinical diagnosis of malignant melanoma of the choroid was made.

SURGERY: On July 23, 1949, an enucleation of the right eyeball was done.

GROSS PATHOLOGY: The specimen is a right eyeball which measures A.P. 24.7 mm., vertically 24.0 mm., and horizontally 24.6 mm. The globe is spherical, of firm consistency, and offers greater resistance to pressure nasally from two to five-thirty o'clock from just anterior to the equator to the optic nerve. The eye transilluminates well except for an area from 2:30 to 5:30 o'clock which does not transilluminate. The cornea is clear and measures vertically 11.7 mm., horizontally 12.8 mm. The iris appears normal. The pupil is circular. The lens gives a metallic sheen to reflected light. The nasal long post ciliary vein is very prominent. The optic nerve is severed flush with the sclera. The greater calotte was taken superiorly in the horizontal plane.

The cornea, iris and lens are in situ and apparently normal. The vitreous cavity is filled with gelatinous, transparent coagulum.

Nasally between the retina and sclera is an irregularly brown mass of firm tissue extending from the ora serrata posteriorly to 4 mm., anterior to the optic nerve and being 6 mm. in width at the equator. The retina is carried into the vitreous cavity over the surface of the neoplasm and is detached below the margin of the neoplasm. Elsewhere the retina is in place. The optic nerve head appears normal and retinal vessels are clearly seen. The macula appears elevated in two places, 1 mm. nasally and 1 mm. temporally to the central dark spot of the fovea centralis in the horizontal plane.

FOLLOW-UP: In May, 1955, the patient was alive and well and there was no evidence of metastases.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 2
December 8, 1957.

ACCESSION NO. 9418
OUTSIDE NO. EIEF 247-55

NAME: R.H.
AGE: 59 SEX: Female RACE: Cau.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient, a fifty-nine year old white female, complained of blurring of vision in the right eye in April, 1955. A field defect was found in the temporal quadrant. The corrected vision however, was 20/25. Ophthalmoscopically there was a large tumor mass in the lower temporal quadrant. A clinical diagnosis of malignant melanoma was made.

SURGERY: On May 1, 1955, an enucleation of the right eyeball was done.

GROSS PATHOLOGY: The specimen is a right eyeball from which the entire cornea has been removed for transplant. The specimen measures A.P. 21 mm., H. 23 mm., and V. 23.5 mm. The sclera is blue-white in color. The optic nerve is 8 mm. long. Transillumination of the globe is good and reveals a shadow approximately 12 mm. in diameter below the insertion of the inferior oblique muscle.

The eyeball was opened obliquely, the larger calotte taken superiority and temporally. The anterior chamber cannot be described. A white lens is in place and is adherent to the posterior surface of the iris. In the inferior temporal quadrant, at the equator, there is a lightly pigmented, pedunculated tumor mass arising in the choroid. The tumor is lobulated and measures 9 x 9 mm. in the region of the large calotte. The retina is detached superiority and nasally, probably artefact of delayed fixation.

FOLLOW-UP: The patient was seen July 3, 1956, for examination and refraction, and again on July 18, 1956, for lens check. At this time the left fundus was normal.
CONTRIBUTOR: A. R. Irvine, Jr., M.D., Estelle Doheny Eye Foundation, 272 South Lake Street, Los Angeles 57, California.

CASE NO. 3

December 8, 1957

ACCESSION NO. 9419
OUTSIDE NO. KDEP 639-56

NAME: E. S.
AGE: 55 SEX: Female RACE: Cauc.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient was thought to have a serous detachment of the retina of the right eye in 1953. In January, 1955, the intra-ocular pressure was 30. A filtering operation was advised, but was not performed. The pressure remained high, but there was no pain. Late in 1955 the eye became red and somewhat painful. A series of Bates exercises was taken without benefit.

The iris changed color and appeared to be replaced by a yellow exudative membrane. At the time of enucleation, December 4, 1956, there were several enlargements of the globe resembling staphylomata. The anterior chamber was filled with a yellow exudate. The globe was hard and blind. At enucleation a thick, yellow mass, presumably neoplasm, attached to the globe over the insertion of the right lateral rectus muscle. This was biopsied. A few days later the globe was enucleated. The left eye was normal.

SURGERY: On December 4, 1956, an enucleation of the right eyeball was done.

GROSS PATHOLOGY: The specimen is a right eyeball measuring A.P. 25 mm., H. 25.5 mm., and V. 24.5 mm. The sclera is white. A firm, slightly pigmented nodule approximately 11 mm. in diameter is present on the temporal aspect over the insertion of the inferior oblique muscle. A similar nodule was removed from the insertion of the lateral rectus muscle at the time of enucleation. The optic nerve is cut flush. The globe does not transilluminate.

The cornea is translucent. The epithelium is intact.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber and the vitreous cavity are filled by a lightly pigmented tumor mass which completely obscures all normal details of the eye.

FOLLOW-UP: The patient expired February 12, 1957. A diagnosis of malignant melanoma of choroid with arteriosclerosis and venous cerebral metastases was made.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 4
December 8, 1957.

ACCESSION NO. 9420
OUTSIDE NO. EDEF 404-51

NAME: H. R.
AGE: 63 SEX: Male RACE: Cau.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient, a sixty-three year old white male, noted scintillating scotomata in the right visual field for three months. A positive scotoma was noted in the nasal field for one month. His vision was 20/40. The lens nucleus was slightly opaque. A black mass that did not transilluminate appeared to extend centrally from the ciliary body between 8 and 11 o'clock to reach the posterior lens capsule just behind the equator at 9 o'clock. The intraocular pressure was normal. A clinical diagnosis of malignant melanoma of the ciliary body was made. The left eye was normal.

SURGERY: On September 12, 1951, an enucleation of the right eyeball was done.

GROSS PATHOLOGY: The specimen is a right eyeball of normal contour measuring A.P. 24 mm., H. 23.5 mm., and V. 23 mm. The sclera is yellow due to fixation. The optic nerve is 2 mm. long. The eyeball does not transilluminate.

The cornea is opaque. A round corneal transplant has been removed from the central portion.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is of normal depth. The iris appears atrophic and two holes are present at 9 o'clock. A small flat lens is in place and is adherent to the posterior surface of the iris. A heavily pigmented tumor mass arises in the ciliary body and choroid temporally. The mass mushrooms into the vitreous cavity and pushes against the posterior lens surface.

The mass measures 10 x 15 mm. and extends from the ciliary body posteriorly past the equator. The retina is separated temporally. A soft mushy opaque coagulum fills the subretinal space.

FOLLOW-UP: The patient is alive. There is no evidence of metastases.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

ACCESSION NO. 9421
OUTSIDE NO. EIEF 354-56

NAME: H. M.
AGE: 52 SEX: Male RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient, a fifty-two year old white male, developed a bullous detachment of the retina in the right eye following an acute episode of posterior scleritis and uveitis. A retinal hole was not found. The retinal elevation subsided from 20 diopters to 5 diopters in a period of a year. The retinal exudates disappeared. The radioactive P32 uptake studies and the Papanicolaou stain of aspirated aqueous were negative in June, 1955. In September, 1955, a cyst-like detachment increased to 20 diopters; in January, 1956, "subretinal fluid was drained from this cyst", and the area was treated with diathermy.

In June, 1956, an acute glaucoma with shallowing of the anterior chamber was noted. The eye was blind. The cornea was edematous. There was perilimbal injection and there appeared to be slight heterochromia. The pupil was dilated and non-reactive. The lens was clear. A cyst-like detachment, rooted in the superior temporal quadrant, was seen in the vitreous cavity. Aspirations of the subretinal fluid revealed cells that were either pigment bearing macrophages or neoplastic cells. The left eye was normal.

The eye was removed because it was painful, blind and glaucomatous. A clinical diagnosis of intra-ocular cyst and/or malignant melanoma was made.

SURGERY: Enucleation of the right eyeball.

GROSS PATHOLOGY: The specimen is a right eyeball of normal contour which measures A-P. 26 mm., H. 25.5 mm., and V. 25.5 mm. The sclera is blue white in color. Numerous round cautery scars are present at the equator on the superior temporal aspect. The optic nerve is approximately 2 mm. long. The globe transilluminates poorly but is firmer superiorly and temporally.

The cornea is translucent. The epithelium is intact.

The globe was opened obliquely, the larger calotte taken temporally and inferiorly. The anterior chamber is shallow. Peripheral anterior synechiae are present. A white lens is present and is displaced anteriorly. The retina is detached between the ora serrata and the optic disc. Inferiorly a firm, gray, opaque coagulum fills the subretinal space. In the superior temporal quadrant the subretinal space is filled with a firm, white mass which contains some pigment. In the region of the large...
calotte the mass protrudes 11 mm. into the vitreous cavity and extends from the ora serrata 11 mm. along the choroid. On the nasal side the mass measures 5 x 11 mm. There is a circular yellow area 6 mm. in diameter on the sclera 14 mm. behind the limbus from 10:30 o'clock.

FOLLOW-UP: In September, 1957, the patient was reported as feeling well and that X-rays of the chest revealed that the chest lesion was unchanged. The referring ophthalmologist expects the patient to report to his office soon.
CONTRIBUTOR: A. R. Irvine, Jr., M.D., Case No. 6A
Estelle Coheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

ACCESSION NO. 9422
OUTSIDE NO. EDEF 122-56

NAME: W. R.  AGE: 53  SEX: Female  RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient was first seen in January, 1956, complaining of loss of central vision in the right eye in the spring and early summer of 1955. A scalloped hemorrhagic area appearing to be about 5 mm. in diameter was seen in and inferior to the macular region. The center of this lesion was already elevated and the edges appeared to be heavily pigmented. Several minute satellite lesions were seen adjacent to the principle mass. The lesions were thought to arise in the choroid because retinal vessels could be seen to course undisturbed over them.

A clinical diagnosis of malignant melanoma was made and the globe was enucleated. The left eye was normal.

SURGERY: On February 6, 1956, an enucleation of the right eyeball was done.

GROSS PATHOLOGY: The specimen is a right eyeball of normal contour measuring A.P. 25 mm., H. 24.5 mm., and V. 24.5 mm. The sclera is white. The optic nerve is approximately 8 mm. long. The globe transilluminates well.

The cornea is transparent. The epithelium is intact.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is of normal depth. A yellow lens is in place and is adherent to the posterior surface of the iris. Strands of a white coagulum extend in the vitreous cavity. The retina is in place. The optic disc is not cupped. The lesion described in the clinical history appears as a white elevated area in the macular area measuring 4 x 8 mm. It has scalloped edges and contains three yellowish cystic-appearing areas. A dark line or indentation can be seen extending from the temporal rim of the disc to the center of the lesion. Grossly there is no evidence of an intraocular neoplasm.

FOLLOW-UP: None.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 6B
December 8, 1957.

ACCESSION NO. 9422
OUTSIDE NO. EDEF 105-49

NAME: M. A.
AGE: 72 SEX: Female RACE:

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient was first seen by the referring ophthalmologist on March 9, 1944, at which time the right eye was blind.

The cornea was opaque and was pigmented brown. The intra-ocular pressure was abnormally high. The anterior chamber was flat. The posterior segment could not be seen. A diagnosis of absolute glaucoma was made and the globe was enucleated. Surgeries of unknown type had been performed upon the cornea five years previously.

SURGERY: Enucleation right eyeball.

GROSS PATHOLOGY: The specimen is a right eye measuring 29.6 mm. antero-posteriorly; 26.7 mm. horizontally; and 25.1 vertically. The anterior half of the eyeball is of normal size, whereas the posterior half is staphylomatous. The cornea measures 14 x 11.3 mm. The cornea is opaque, and a central band-shaped opacity is present. There is perilimbal injection.

The eye was opened horizontally. The anterior chamber is flat. The lens is large and cataractous. There is marked atrophy of the choroid and retina posteriorly. Three millimeters temporal to the disc in the horizontal meridian is a yellow, elevated mass sharply circumscribed and about 3 mm. in diameter.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.
CASE NO. 68

ACCESSION NO. 9422
OUTSIDE NO. EDEF 523-49

NAME: L. J.
AGE: 60 SEX: Female RACE: White

Tissue FROM: Right eyeball.

CLINICAL FINDINGS: This patient complained of blurring central vision in the right eye. One week after the onset of symptoms the vision in the right eye was 20/200. A circumscribed mass was seen in the macula and contained multiple brown-whitish elevations, the advancing edge of which appeared to be hemorrhagic. A clinical diagnosis of a rapidly growing malignant melanoma was made.

SURGERY: Enucleation of right eyeball.

GROSS PATHOLOGY: The specimen is an enucleated right eyeball measuring 23 mm. x 24 mm. x 24 mm., firm to pressure throughout and of average consistency. The optic nerve stump measures 2 mm. Two black silk sutures are attached to the globe at the insertions of the medial and lateral recti muscles. The eyeball transilluminates well throughout.

The cornea is clear. There is a whitish band encircling almost the entire cornea just within the limbus leaving a clear area between it and the limbus. There is a tiny opacity centrally in the cornea. The anterior chamber is moderately deep. The pupil is regular and measures 4 mm. in diameter. The lens appears cataractous through the pupillary opening.

The globe was sectioned horizontally above, the major calotte being taken above. The lens is greenish-yellow and there are streaks along its posterior surface. The lens appears large and appears to be pushing the iris diaphragm forward. The anterior chamber is shallower than was thought prior to opening the globe. The vitreous is clear and of a semi-solid consistency.

The significant finding is in the posterior pole.

Extending temporally from the disk is a round, flat, but slightly elevated area which is darker than the surrounding retina and choroid. This area includes the macula and measures 8 mm. in diameter. The retina is thrown into a thickened grayish fold which passes over this area horizontally and another fold which passes obliquely to the horizontal fold. The retinal vessels pass over the medial aspect of this elevated area. The retina appears thickened in the region of the disk. The vessels are well visualized and are partially covered in the region of the disk. The optic disk is not cupped. The retina is adherent throughout.

FOLLOW-UP: None.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.  
Estelle Doheny Eye Foundation  
272 South Lake Street,  
Los Angeles 57, California.  

CASE NO. 7  
December 8, 1957

ACCESSION NO. 9423  
OUTSIDE NO. MDEF 433-34

NAME: P. E.  
AGE: 72  
SEX: Male  
RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: This patient has had a growth in the anterior chamber of the right eye known to be present for ten years. Eight years after its onset it was approximately 2 mm. in diameter and attached to the free margin of the iris. It increased rapidly in size over the following twenty-one months and at the time of enucleation it occupied the inferior two-thirds of the anterior chamber. The corrected vision was reduced to 20/100. There was no evidence of glaucoma. A clinical diagnosis of malignant melanoma of the iris was made.

SURGERY: Enucleation of right eyeball.

GROSS PATHOLOGY: The specimen is a right eyeball of normal size and shape. The cornea is clear. There is a chocolate brown mass apparently arising from and adherent to the iris inferiorly, partially filling the anterior chamber and obliterating the pupil in an oblique line extending from 10 to 3:30 o'clock. The pupil above is dilated and transilluminates normally.

The eyeball was opened horizontally, the larger calotte taken below. A black tumor fills the anterior chamber below. The rest of the specimen is not remarkable.

FOLLOW-UP: None.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 8
December 8, 1957.

ACCESSION NO. 9424
OUTSIDE NO. BDEF 268-50

NAME: C. P.
AGE: 49 SEX: Female RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: This patient was first seen in October, 1949, presenting the findings of acute congested glaucoma. Both irides were blue, the intraocular pressure was reduced with medication. A basal iridectomy was performed.

The pressure remained normal and the patient remained asymptomatic until January, 1950, at which time the right iris appeared to be infiltrated with black pigmented tissue apparently arising from the iris angle below. The tension became elevated, the eye painful and congested, and the cornea edematous. A clinical diagnosis of malignant melanoma was made. The left eyeball also presented a shallow anterior chamber and chamber angle and subsequently developed acute narrow angle primary glaucoma.

SURGERY: Enucleation of right eyeball.

GROSS PATHOLOGY: The specimen is an enucleated right eyeball which measures A.P. 23 mm., H. 22.5 mm., and V. 23 mm. The sclera has a bluish tint in the area of the ciliary body. There is a total coloboma of the iris from 10 to 12:30 o'clock. A heavily pigmented mass arising from the iris fills the anterior chamber. The eyeball transilluminates well except in the area of the ciliary body. The upper larger calotte was removed. The tumor invades only the iris and the ciliary body; the remaining structures of the eye show no gross evidence of tumor. The lens and retina are in place. The iris is adherent to the lens, and there is moderate cupping of the optic disc.

FOLLOW-UP: The patient was last seen in 1954 and was in good condition. She has moved to Northern California.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 9 and
CASE NO. 10.

December 8, 1957.

ACCESSION NO. 9425 and 9426.
OUTSIDE NO. EDEF 257-50 and EDEF 5-57.

NAME: A. W.
AGE: 47 SEX: Female RACE: Cauc.

TISSUE FROM: Epibulbar tissue.

CLINICAL FINDINGS: In 1944, at the age of forty-seven, this patient developed an epibulbar tumor in the left eye. The tumor measured 4 x 7 mm. in diameter and was located in and beneath the bulbar conjunctiva adjacent to the limbus inferiorly. It was resected. On six occasions during a subsequent thirteen year period similar epibulbar tumors were removed from this eye. The first developed five years after the original tumor was resected and was approximately in the same location. There was no evidence of metastases. The family history was of significance in that an older sister had an eye removed because of a choroidal melanoma.

Because of the five year interval since the onset of symptoms it was considered that if the tumor were to metastasize it would have done so, and therefore a local wide excision of the lesion was done rather than an exenteration. New tumors were seen and resected in 1951, 1954 and 1955. Each time the neoplasm was resected widely and the margins of the excised tissue found to be normal microscopically. In January, 1957, a swelling of the lower lid margin developed. An exenteration was performed.

GROSS PATHOLOGY: The exenterated specimen of the left orbit measures A.P. 39 mm., H. 32 mm., and V. 21 mm. Portions of the upper and lower lid were removed for biopsy.

The cornea is clear. The epithelium is intact. A crescent shaped elevation is present on the conjunctiva inferiorly, the site of previous mucous membrane transplant.

The eyeball was opened obliquely, the larger calotte taken superiorly and temporally. The anterior chamber was of normal depth. A large, white lens was in place. Strands of a white coagulum extended into the vitreous cavity. The retina was separated posteriorly, probably artefact.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.

Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 11
December 8, 1957

ACCSSION NO. 9551
OUTSIDE NO. EDEF 369-52

NAME: C. H.
AGE: 58 SEX: Male RACE: Caucasian

TISSUE FROM: Right eyeball

CLINICAL FINDINGS: This patient complained of symptoms of prostatism for a year, and generalized weakness and anemia progressive for six months. For two weeks his vision had been blurred and experienced diplopia when looking to the right. Many tumorous masses were seen on the skull a few months prior to death.

The vision in the right eye was reduced to hand movements; in the left eye, to counting fingers. The right pupil was 3 mm. in diameter and reacted normally, whereas the pupil of the left eye was 6 mm. in diameter and was fixed. A large hemorrhage was seen on the nasal side of the disc. Some exudates were present. A small hemorrhage was seen below the left disc. The eye was removed at autopsy.

SURGERY: Enucleation of right eyeball August 3, 1952.

GROSS PATHOLOGY: The specimen is a right eyeball of normal contour measuring A.P. 23 mm., H. 24 mm., and V. 23 mm. The sclera is blue white in color. The optic nerve is cut flush with the sclera. Transillumination of the eyeball is good.

The cornea is translucent, with a narrow, white, opaque area at the limbus between 7 and 9 o'clock. The epithelium is intact, but is wrinkled, probably artefact.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is shallow. A white lens is in place and is adherent to the posterior surface of the iris. A translucent coagulum fills the vitreous cavity. The retina is in place, with numerous folds radiating from the optic disc. The macular area is white and elevated.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.  
Estelle Doheny Eye Foundation,  
272 South Lake Street,  
Los Angeles 57, California.  
CASE NO. 12  
December 8, 1957

ACCESSION NO. 9478  
OUTSIDE NO. EDEF 210-48

NAME: E. C.  

TISSUE FROM: Caruncle.

CLINICAL FINDINGS: The patient has had a pigmented lesion of the left caruncle for many years. Recently it has increased in size and has become more vascular. It appeared to be somewhat cystic and slightly elevated. A clinical diagnosis of either a nevus or a malignant melanoma of the caruncle was made.

* * * * * * * * * * * * * * * * *

CONTRIBUTOR: A. R. Irvine, Jr., M.D.  
Estelle Doheny Eye Foundation,  
Los Angeles 57, California.  
CASE NO. 12A  
December 8, 1957

ACCESSION NO. 9478  
OUTSIDE NO. EDEF 528-49

NAME: H. B.  
AGE: 53 SEX: Male RACE: White

TISSUE FROM: Conjunctiva from caruncle.

CLINICAL FINDINGS: The patient complained of a "mole" at the inner angle of the right eye. It had been present for two years and had apparently increased in size.

The eyes were normal except for a pigmented nevus of the right caruncle. The pigmentation extended into the fornix and palpebral conjunctiva in the region of the punctum below. Temporally it extended about one millimeter into the bulbar conjunctiva. It was easily undermined and excised under local anesthesia.

FOLLOW-UP: The patient was last seen in 1951 at which time there was no evidence of recurrence.

The sections studied indicated that the lesion was incompletely removed.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.
CASE NO. 128
December 8, 1957

ACCESSION NO. 9478
OUTSIDE NO. EDEF 503-52

NAME: P. P.
AGE: 8 SEX: Male RACE: Cau.

Tissue From: Bulbar conjunctiva.

CLINICAL FINDINGS: This child had had a pigmented lesion of the bulbar conjunctiva for several years. It was removed because it was increasing in size. A clinical diagnosis of conjunctival nevus was made.

* * * * * * * * * * * * * * * *

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.
CASE NO. 128

ACCESSION NO. 9478
OUTSIDE NO. EDEF 346-52

NAME: W. B. (Mrs.)
AGE: 31 SEX: Female RACE: White

Tissue From: Conjunctiva.

CLINICAL FINDINGS: The patient first noticed a brownish pigmented tumor of the temporal bulbar conjunctiva adjacent to the limbus when she was thirty years of age. It appeared to have increased in size. The tumor was slightly elevated and appeared to contain minute cysts. Another pigmented lesion considerably smaller and flatter was seen adjacent to the nasal limbus. This had been present since early childhood and had not changed in appearance. The larger lesion was removed for cosmetic reasons. A diagnosis of conjunctival nevus was made.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Dorothy Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 13
December 8, 1957.

ACCISSION NO. 9427
OUTSIDE NO. EDEF 512-56.

NAME: M. J.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: The patient was found to have conjunctivitis in the right eye in May, 1956, at which time an antibiotic was prescribed. Since that time the parents occasionally noticed a white reflex in the right pupil. In September, the father determined that the patient could not see out of the right eye.

The examination at that time indicated a yellowish fundus reflex. The globe was of normal size. The anterior chamber was of normal depth. Particulate material could not be seen floating in the anterior chamber or in the vitreous, but small whitish deposits on a yellowish tumor behind the lens could be seen. X-rays for calcium were negative. The optic foramen was not enlarged. A clinical diagnosis of retinoblastoma was made. The other eye was normal on examination under ether.

SURGERY: Enucleation of right eyeball performed on September 25, 1956.

GROSS PATHOLOGY: The specimen is a right eyeball which measures A.P. 25 mm., H. 23 mm., and V. 21.5 mm. The sclera is blue-white in color. The optic nerve is approximately 7 mm. long. The globe does not transilluminate, however, it is quite firm temporally.

The cornea is translucent. The epithelium is intact.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is very shallow. Peripheral anterior synechiae are present. A yellow lens is in place. The retina is in place. The temporal half of the vitreous cavity is filled with a friable white tumor mass containing small areas of calcification. Nasally the vitreous cavity is filled with a white opaque coagulum. In the posterior temporal quadrant and also inferiorly a firm, white mass separates the retina and choroid. This area measures 2 x 16 mm. in the region of the large calotte. Tumor seedlings can be seen along the retina nasally.

FOLLOW-UP: Three months postoperatively the tumor recurred in the orbit. The extent of the metastasis was to the head, bones of head. The patient expired in August, 1957.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 14

December 8, 1957

ACCUSION NO. 604
OUTSIDE NO. B.H.E. 230-49

NAME: P. B.
AGE: 18 months SEX: Male

TISSUE FROM: Left eyeball

CLINICAL FINDINGS: The patient was seen five months after the parents noticed that his left eye had gradually changed from blue to brown in color.

A tumor mass was seen behind the iris and anterior to the lens. The iris appeared grossly vascular and atrophic over the tumor. On transillumination, the tumor appeared to be confined to the temporal iris in the 3 o'clock meridian. The intra-ocular pressure was elevated to 56 mm. Hg (Schiotz). A clinical diagnosis of "intra-ocular tumor" was made.

SURGERY: On May 18, 1949, an enucleation of the left eyeball was performed.

GROSS PATHOLOGY: The specimen consists of an enucleated left eyeball which is uniformly normal in consistency and normal in shape. The sclera is a little more bluish than average. The eyeball measures 23 mm. A.P., 23 mm. vertically, and 23 mm. horizontally. It transilluminates well from all points posteriorly, but there is an elliptical shadow 1 mm. in width extending from 12:30 to 2:30 o'clock at the pupillary margin. It gives the appearance of a growth extending inward between the lens and the iris. The cornea is transparent and measures 12 mm. vertically and 13 mm. horizontally. The anterior chamber is approximately 3 mm. in depth. The iris is broader from 2:30 to 6:00 o'clock than is its remaining portion. That part just above 2:30 o'clock shows a small membrane which is free at one margin. Peripheral to this membrane there is a thinning of the iris, or its pigment, to produce a light, gray-colored, elliptical area with brown stippling. The pupil is not distorted. The optic nerve was severed 1.5 mm. from the sclera and measures 3.5 mm. with its coverings.

The greater calotte was taken above about 20 degrees from the horizontal meridian to expose, but not disturb, the above area. Transillumination after opening shows a slight thickening of the ciliary body and a disturbance in the normal pattern of the corona ciliaris in the area previously outlined. This area readily transilluminates but not as well as adjacent similar structures. There is a grayish white membrane covering the ciliary body and extending slightly onto the lens at this point. The lens is not grossly displaced and is semi-transparent. The vitreous chamber is filled with a semi-transparent jelly-like homogeneous substance. The retina and choroid are intact. The optic disc is normal and the sclera is slightly thinner than average.
CONTRIBUTOR: Leonard Christensen, M.D.
University of Oregon
Medical School,
Portland 1, Oregon.

ACCESSION NO. 604
OUTSIDE NO. 947

NAME: M. M.
AGE: 3 yrs. SEX: Female RACE: Cau.

TISSUE FROM: Left eyeball.

CLINICAL FINDINGS: This specimen was obtained from a three year old white female in January, 1954. The presenting complaint was a painful, injected left eye which had been tender for the previous three months. During this period the eye had a tendency to turn up and out and she rubbed it constantly. Examination showed a shallow anterior chamber and a dilated, irregular pupil. There was photophobia, tenderness and constant tearing. The cornea was bedewed and tonometer reading (Schiotz) was 46 to 49 mm. of mercury. Funduscopy after clearing the cornea revealed a complete funnel-shaped retinal separation. No normal retina could be distinguished even after dilating the pupil. X-ray examination for calcium was negative.

SURGERY: Enucleation of left eyeball.

GROSS PATHOLOGY: The specimen was firm and a regularly shaped left eye measuring 24 mm. in the AP diameter by 24 mm transversely at the equator.

The cornea was clear showing a moderately deep anterior chamber which was free of cells and debris. The pupil was widely dilated, round and regular. There was minimal perilimbal neovascularization. The remainder of the external examination showed nothing of note. Transillumination demonstrated a solid, opaque mass in the inferior temporal quadrant.

Inferior horizontal calotte: On opening a moderate amount of clear, subretinal fluid escaped. The retina was detached to a central strand with an umbrella-like dome behind the lens. Lying between the retina and temporal choroid was a large, solid mass showing numerous hemorrhagic areas on the cut surface. Grossly, no definite origin of the tumor could be determined since it was closely adherent to the choroid as well as the retina. The nerve head was not seen because of complete detachment of the retina.

Paraffin sections of the optic nerve were negative for neoplasm.

CASE NO. 15

December 8, 1957

ACCESSION NO. 9428
OUTSIDE NO. PDEF 501-56

NAME: R. R.

TISSUE FROM: Right eyeball

CLINICAL FINDINGS: This patient was noted to have a white mass behind the lens of the right eye three days after birth. The anterior segment appeared normal. The lens seemed to be slightly luxated to the temporal side. The pupil was slightly irregular but reactive. The cornea was clear. A white stellate mass was seen in the anterior vitreous and normal vessels passed through it. The posterior segment of the globe could not be visualized. The left eye was normal.

The mother of the patient had toxemia at pregnancy. The child was born at full term and was jaundiced at birth. X-rays of the optic foramen were normal and there were no apparent calcifications. The anterior chamber became shallow and the cornea became ectatic. A tentative clinical diagnosis of congenital anomaly, granuloma, or retinoblastoma was made.

SURGERY: Emucleation of the right eye, September 17, 1956.

GROSS PATHOLOGY: The specimen is an elongated right eyeball measuring A.P. 22 mm., H. 18 mm., and V. 18 mm. The sclera is blue. The optic nerve is approximately 3 mm. long. Transillumination of the eyeball is good.

The cornea is translucent, is enlarged and measures 13 x 13 mm. The epithelium is intact. The pupil is elliptical with vertical axis and is displaced toward 9 o'clock.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is absent. The iris is divided, the anterior section forming an anterior synechia on the bulging cornea and encircling a small lens which adheres to the posterior surface of the cornea. The lens is displaced temporally. There is a defect in the lens posteriorly. The posterior half of the iris is in its normal position. A small section of lens fills the pupil. A thin, white membrane extends across the posterior surface of the iris. A translucent coagulum fills the anterior half of the vitreous cavity and extends in a single strand and attaches to the nasal rim of the optic disc. The retina is in place. The optic disc is cupped.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.  CASE NO. 16
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.
December 8, 1957

ACCESSION NO. 9429
OUTSIDE NO. EDEF 160-55

NAME: J. A.
AGE: 2½ SEX: Male RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: This patient was noted to have an intra-ocular tumor in the right eye since the age of eight months. Exotropia was the presenting sign. The child remained untreated because the attending oculist advised the parents that the lesion was benign.

At examination a large retinal detachment was seen in the right eye and a tumor mass was seen filling the inferior half of the vitreous cavity. X-rays of the orbits and optic foramina were normal. A clinical diagnosis of retinoblastoma was made.

SURGERY: Enucleation of right eyeball.

GROSS PATHOLOGY: The specimen is a small right eyeball measuring A.P. 21 mm., H. 21 mm., and V. 19.5 mm. The sclera is blue white in color. The optic nerve is cut flush. The eyeball does not transilluminate.

The cornea is transparent. The epithelium is intact.

The eyeball was opened vertically, the larger calotte taken temporally. The anterior chamber is deep. A thin flat lens is in place. A white membrane extends from the inferior ciliary body up across the posterior lens surface and to the equator superiorly. It appears that the retina is completely detached between the ora serrata and the optic disc. A firm, tan, translucent coagulum fills the subretinal space. A small, round, brownish area approximately 3 mm. in diameter is present superiorly at the attachment of the white membrane.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 17.
December 8, 1957

ACCESSION NO. 9430
OUTSIDE NO. EDEF 331-49

NAME: J. C. R.
AGE: 76 SEX: Male RACE: White

TISSUE FROM: Left eyeball.

CLINICAL FINDINGS: This patient was known to have a growth involving the left cornea and bulbar conjunctiva for twelve years. Although the lesion increased in size, the patient refused enucleation until persistent discharge and severe pain developed. At the time of the enucleation he was unable to close his lids. A cauliflower mass covering the lower three-fourths of the cornea and measuring 25 mm. H. and 8-15 mm. V. was seen. The cornea was not involved above. The posterior segment could not be viewed. A clinical diagnosis of epithelioma of the limbus was made.

SURGERY: Enucleation of left eyeball.

GROSS PATHOLOGY: The specimen consists of a left eyeball measuring 23 mm. anteroposteriorly, 22 mm. horizontally and 21 mm. vertically. There is a lobulated white meaty tumor mass covering all but the superior third of the cornea. It extends from the limbus nasally as a white elevated band across the cornea to about 4 mm. posterior to the temporal limbus. Its maximum measurements are 21 mm. horizontally, 11 mm. vertically and 10 mm. anteroposteriorly. Palpation of the eyeball does not reveal any abnormal firmness other than in the area of the tumor mass. The eye transilluminates normally through the superior clear quarter of the cornea.

The eye was opened horizontally, the greater calotte taken above. The vitreous cavity is filled with a gelatinous translucent coagulum. The interior of the eye is essentially normal.
CONTRIBUTOR: A. R. Irvine, Jr., M.D. CASE NO. 18
Estelle Doheny Eye Foundation, December 8, 1957
272 South Lake Street,
Los Angeles 57, California.

ACCESSION NO. 9431
OUTSIDE NO. EDEF 188-57

NAME: A. C.
AGE: 66 SEX: Female RACE: Caucasian.

TISSUE FROM: Bulbar conjunctiva.

CLINICAL FINDINGS: This patient has had two previous operations for a bulbar conjunctival lesion described as a squamous cell carcinoma. It has recurred on each occasion and for the past three months has invaded the anterior fifth of the cornea temporally in the left eye. A piece was removed and studied in frozen section and thought to represent Bowen's dyskeratosis. The cornea was peeled temporally. The involved conjunctiva was excised and the defect covered by undermining conjunctiva and approximating the severed edges.

* * * * * * * *

CONTRIBUTOR: A. R. Irvine, Jr., M.D. CASE NO. 18A
Estelle Doheny Eye Foundation, Los Angeles 57, California.
272 South Lake Street,

ACCESSION NO. 9431
OUTSIDE NO. EDEF 30-56

NAME: C. D.
AGE: 32 SEX: Male RACE: Caucasian.

TISSUE FROM: Limbal conjunctiva.

CLINICAL FINDINGS: The patient noticed a growth involving the nasal limbal conjunctiva in the right eye in December, 1955. It increased in size rapidly and at the time of removal it was about 4 mm. in diameter and was elevated and non-pigmented. It was moderately vascular and was accompanied by considerable bulbar injection. A clinical diagnosis of possible Bowen's disease of the limbus was made.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

ACCESSION NO. 9431
OUTSIDE NO. EDEF 65-56

NAME: G. L.
AGE: 27 SEX: Male RACE: White

TISSUE FROM: Bulbar conjunctiva.

CLINICAL FINDINGS: The patient was first seen January 23, 1956, at which time he had noticed a small nodule on the lateral aspect of the left eye for several weeks. Clinically this nodule appeared as a flesh colored mass measuring 4 mm. to 5 mm. in diameter, involving the conjunctiva near the limbus. A few large vessels were fanning out from the nodule. The patient was placed on topical cortisone for one week with little or no response. His vision was 20/15 in each eye.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 19
December 8, 1957

ACCUSSION NO. 9432
OUTSIDE NO. EDEF 165-51

NAME: L. T. S.
AGE: 57 SEX: Female RACE: White

TISSUE FROM: Portion of excised iris.

CLINICAL FINDINGS: This patient was found to have a tumor of the left iris during a routine examination. A small, brownish, milky appearing lesion involving the iris between the 1 and 3 o'clock meridian was seen. The normal contour of the pupil became flattened under mydriasis. The surface of the neoplasm appeared to be regular. There were small tortuous blood vessels that dipped into the normal surrounding blue iris stroma. Gonioscopically the chamber angle was uninvolved. The intra-ocular pressure was normal. A clinical diagnosis of malignant melanoma or leiomyoma of the iris was made. A biopsy was performed.
The patient was seen in consultation on January 2, 1941, for an opinion as to the nature of a small growth on the iris of the right eye. The growth had been apparent for about two years, but only recently seemed to be getting larger. There had been no symptoms. The vision was 20/20 with correction; the eye appeared white and quiet, there being no evidence of inflammation observed on slit lamp examination; the media were clear; the fundus was normal.

On examination, a mulberry-shaped, cystic appearing grayish, sessile tumor, 3 mm. in diameter, was seen resting on the iris in the lower quadrant midway between the pupillary margin and the iridocorneal angle. Each mulberry papilla contained a capillary tuft. The pupil reacted well; however, there was slight pulling of the pupillary edge toward the growth. The adjacent iris stroma on all sides appeared normal. The mass did not extend into the iridocorneal angle. Because of noticeable growth during the past year, iridectomy for removal of the tumor was advised.

On January 10, 1941, an uncomplicated basal iridectomy was performed, the growth being removed along with a good margin of iris. Although the growth appeared cystic and soft, when grasped with the iridectomy forceps it felt hard and resistant.

The lesion consisted of a rounded sessile mass 3 mm. in diameter situated on the anterior surface of the iris. The tumor was white and semi-translucent. Its surface was smooth and glistening.

The patient made an uneventful recovery, maintaining 20/20 vision, and up to sixteen months after operation, the eye has remained normal except for the operative coloboma.
CONTRIBUTOR: Ralph H. Fuller, M.D.,
St. Mary's Hospital,
Tucson, Arizona.

CASE NO. 19B
December 8, 1957

ACCESSION NO. 9432
OUTSIDE NO. EDEF 368-55

NAME: F. C.
AGE: 45 SEX: Male RACE: White

TISSUE FROM: Iris biopsy.

CLINICAL FINDINGS: The patient was first seen in May, 1955, at which time he was found to have an inflamed pinguecula. A month later a grayish-yellow elevation was seen on the iris at 8 o'clock. A few small blood vessels were apparent and a few hemorrhagic extravasations. It measured about 2 x 3 cm., and was elevated a millimeter above the surface of the iris. There appeared to be normal iris tissue surrounding the tumor. In June, 1955, the lesion was excised under local anesthesia.

FOLLOW-UP: When last seen in April, 1956, there was no evidence of recurrence.
The patient developed progressive proptosis of the right eye over a six year period. Two years after the onset, surgical exploration failed to reveal the cause. In July, 1956, a glioma the size of a walnut was found. This and the globe were removed retrograde through the frontal approach.

The globe was blind and at the time of removal the nerve was atrophic and projected into a slant. A clinical diagnosis of glioma was made. The left eye was normal.

The specimen is a Zenker fixed right eyeball measuring A.P. 21 mm., H. 23 mm., and V. 23 mm. The sclera is brown. The optic nerve is approximately 4 mm. long. The distal end of the nerve is much broader than normal, measuring H. 8 mm., and V. 9 mm. The actual nerve tissue appears to be split horizontally. The globe does not transilluminate. Two pieces of tumor tissue are enclosed with the eyeball; one measures 22 x 20 x 5 mm., the other 8 x 20 x 5 mm.

The cornea is opaque. The epithelium is denuded in two small areas centrally.

The eyeball was opened horizontally, the larger calotte taken superiorly. The anterior chamber is of normal depth. A white lens is in place and is adherent to the posterior surface of the iris. A soft, yellow coagulum fills the vitreous cavity. The retina is separated, probable artefact of fixation.
CONTRIBUTOR: Dennis S. Shillam, M.D.  
Huntington Memorial Hospital,  
100 Congress Street,  
Pasadena 2, California.  

CASE NO. 21  
December 8, 1957.  

ACCESSION NO. 9434  
OUTSIDE NO. 81791-55  

NAME: N. K. Z.  
AGE: 52  SEX: Female  RACE: White  

TISSUE FROM: Tumor, right orbit.  

CLINICAL FINDINGS: The patient was admitted to the hospital on May 2, 1955, giving a history of the appearance of a small mass in the upper outer quadrant of the right orbit in 1953. A biopsy of the mass in 1954 was reported as normal lacrimal gland. There was a steady increase in the size of the mass with definite ptosis of the right eyelid. Contrast orbital X-rays revealed a tumor mass in the region of the right lacrimal gland. The only symptom was occasional diplopia in the morning.

The physical examination showed moderate proptosis of the right eye, with a palpable mass involving the upper outer quadrant of the orbit, causing the globe to be pushed downward, inward and outward. The past history included tonsillectomy and adenoidectomy approximately at the age of 30, elsewhere; thyroidectomy for thyroiditis in 1947; and hysterectomy for leiomyomata in 1950. On May 3, 1955, a right frontal bone flap craniotomy was performed and the orbit entered through the roof, which was found to be extremely thin. A tumor in the right lateral superior angle of the orbit was dissected out by blunt and sharp dissection from the orbital tissues, and was felt to lie predominantly external to most of the orbital contents and was compressing them downward and forward. This large tumor was connected by a small neck to the region of the right lacrimal gland.

GROSS PATHOLOGY: The specimen received in the laboratory was red-yellow tissue measuring 3.5 x 2.5 x 1.8 cm. At one end was a suture around a cord-like structure 4 mm. in circumference. Sections exposed soft, gray-white tissue occurring in multiple small nodules from 3 to 8 mm. in diameter.

FOLLOW-UP: The patient was last seen by the Neurosurgical consultants in June, 1955. In 1956, a lymph node developed in the right supraclavicular region.
CONTRIBUTOR: Armed Forces Institute of Pathology, Walter Reed Army Medical Center, 6825 16th Street, NW., Washington 25, D.C.

CASE NO. 22
December 8, 1957.

ACCESSION NO. 9435
OUTSIDE NO. AFIP No. 677710

AGE: 12 SEX: Female RACE: White

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS: This patient complained of pain and swelling of the right eye. She was presumably in good health until five months prior to admission when she noted frontal headaches followed by difficulty in reading print. Three weeks before admission, intermittent swelling of the right eye was noted; this became persistent four days prior to admission. There was slight pain and photophobia. There was no discharge from the eye or nose. The visual acuity remained normal.

The pupil was dilated. There was marked exophthalmus, edema of the lid and congestion of the globe. The extraocular movements were limited and the nerve was edematous. The pupils were normally reactive. The visual fields were full. The neurologic examinations were negative.

There was a mild anemia. The spinal fluid was normal. X-rays of the chest, spine, pelvis and femora were negative. X-rays of the skull revealed some erosion of the anterior portion of the sphenoid ridge of the left side while it appeared to be intact on the right.

A neurosurgical examination was done under pentothal anesthesia. An orbital mass was removed with the globe. A pre-operative diagnosis of neurofibroma or abscess of nerve was made.

GROSS PATHOLOGY: The specimen received in the laboratory consisted of a collapsed eye with the cornea completely inverted into the globe. A large indurated mass measuring 39 x 37 x 17 mm. was attached to the posterior and inferior aspects of the globe. This apparently represented only a portion of the mass that had been transected, the other portion of the mass apparently having been retained in the local hospital laboratory. The transected optic nerve could be identified within the center of this mass and portions of rectus muscles could be seen about its periphery. The indurated tissue presented a diffuse pale silvery-gray appearance with yellowish mottling.
CASE NO. 23
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

December 8, 1957.

ACCESSION NO. 9436
OUTSIDE NO. ERF 527-54

NAME: F. C. L.
AGE: 60 SEX: Male RACE: White

TISSUE FROM: Left eyeball

CLINICAL FINDINGS: This patient entered the hospital for the second and last time on August 30, 1954, because of a large mass in the abdomen, swelling in the legs and progressive weakness. For forty years he had nodules beneath the skin of the lower thorax and upper abdomen. In 1951, a biopsy of one of these skin nodules was reported as neurofibromatosis. In 1953, a biopsy of another nodule was reported as a benign subcutaneous fibroma.

In July, 1951, he noticed a scotoma in the left eye. The retina was found to be detached below the optic disc. Diathermy surgery for retinal detachment was done in June, 1953. The retinal detachment clinically resembled a cyst and the detachment enlarged so that it occupied most of the inferior half of the vitreous cavity and obscured the interior two-thirds of the disc. Diathermy and trephination of the sclera was done in January, 1954, without success.

At the time of examination in October, 1954, X-ray revealed densities in the left lung field. These were typical metastatic lesions. The abdomen was distended, the liver enlarged and nodular, and the legs were edematous to the knees. The hospital course was progressively downhill with increase in weakness, weight loss and edema. The patient expired October 23, 1954.

GROSS PATHOLOGY: The specimen is a Zenker fixed left eyeball measuring A.P. 22 mm., H. 25 mm., and V. 26.5 mm. The sclera is yellow due to Zenker fixation. The optic nerve is 13 mm. long. The eyeball does not transilluminate.

The cornea is opaque. The epithelium is wrinkled.

The eyeball was opened obliquely, the larger calotte taken superiorly and nasally. The anterior chamber is shallow. A round yellow lens is in place. A mushy yellow coagulum fills the vitreous cavity. The retina is separated posteriorly and the sub-retinal space is filled with a firm yellow substance.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

CASE NO. 24
December 8, 1957.

ACCESSION NO. 9437
OUTSIDE NO. 1111-56

NAME: G. L. R.
AGE: 87 SEX: Female RACE: Cauc.

TISSUE FROM: Right eyeball.

CLINICAL FINDINGS:
In February, 1954, this patient had an extracapsular cataract extraction performed on the right eye. The eye remained painful and inflamed postoperatively despite the use of steroids and mydriatics.

Three years later at the time of enucleation the vision was reduced to light perception. The cornea was edematous and the perilimbal area was injected. There was a positive aqueous flare. The iris was incarcerated in the wound at 10 o'clock. A thick pupillary membrane was noted and obscured the view of the posterior segment. The intraocular pressure was 60 units. A diagnosis of uveitis secondary to cataract surgery was made. There was a nuclear cataract in the left eye.

SURGERY:
On May 9, 1956, an enucleation of the right eyeball was performed.

GROSS PATHOLOGY:
The specimen is a right eyeball which measures A.P. 25 mm., H. 24 mm., and V. 24.5 mm. The sclera is blue-white in color and is thinned superiorly and temporally at the equator. The optic nerve is cut flush. The globe transilluminates well.

The cornea is opaque centrally and translucent at the periphery between 2 and 10 o'clock. The epithelium is denuded at 9 o'clock. A linear scar extends at the limbus between 10 and 2 o'clock.

The eyeball was opened vertically, the larger calotte taken temporally. The anterior chamber is absent. There is an anterior synechia. Lens remnants are adherent to the posterior surface of the iris. A white coagulum fills the vitreous cavity. The retina is in place. Details of the optic disc are obscured by the coagulum.
CONTRIBUTOR: Albert F. Brown, M.D., Glendale Sanitarium and Hospital, 1509 East Wilson Avenue, Glendale 6, California. CASE NO. 25

ACCESSION NO. 9395
OUTSIDE NO. GSH 57-1468

NAME: A. J. D.
AGE: 38 SEX: Male

TISSUE FROM: Contents of right orbit including the eyeball and eyelids, an irregular 5 x 4.5 x 3 cm. piece of bone, and portions of muscles.

CLINICAL FINDINGS: This patient complained of pain and numbness in the right side of his face (upper lip and below eye) for five months. He had a past history of sinus trouble, asthma and postnasal discharge. The left antrum was operated on fifteen years ago and opaque media introduced in both sinuses for X-rays. Three months ago the right antrum was explored and residual opaque material removed. Tissue obtained was reported as benign. Subsequently, he noticed a small swelling in the lower eyelid, displacing the eye upward. Biopsy of this swelling was performed.

In April, 1957, the patient was admitted again and for definitive surgical treatment, which consisted of exenteration of the right orbit and partial resection of the superior maxilla on the right. At operation, a firm mass was found beneath the inner third of the right orbit which was firm and nontender. On reflecting the facial soft tissues the tumor mass was found to occupy the inner portion of the orbit, the right lateral wall of the nose superiorly, and the ethmoidal sinuses.

GROSS PATHOLOGY: The specimen consists of the contents of the right orbit including the eyeball and eyelids, an irregular 5 x 4.5 x 3 cm. piece of bone, and portions of muscles. The skin attached to the lower eyelid is 2.5 cm. in greatest width. At the lower anterior medial angle there is an irregular 2 cm. firm, yellow-white, poorly encapsulated mass. This does not grossly appear in the adjacent conjunctival surfaces or surfaces of the eye. Also present is a separate 9 x 2 x 0.8 cm. piece of adipose tissue, many irregular flat pieces of bone up to 4.5 x 3 x 0.5 cm., a 4.5 x 3 x 1 cm. piece of mucous membrane with some associated fragments of bone, a 15 x 4 mm. segment of apparent nerve tissue, a moderate number of small pieces of bone up to 1 cm. long which together form a 2 cm. mass, and many pieces of mucous membrane with associated small pieces of bone up to 2.5 cm. in length.

FOLLOW-UP: The patient was discharged May 13, 1957. He re-entered July 8th, 1957, for postoperative fistula of right cheek. No residual tumor was found at this time.
ADDENDA

TO THE

CALIFORNIA CANCER COMMISSION
SEMI-ANNUAL SLIDE CONFERENCE

ON

NEOPLASMS OF THE EYE AND ADNEXA

December 8, 1957

A. Dr. Irvine's diagnosis.
B. Histology and discussion.
C. Bibliography.
<table>
<thead>
<tr>
<th>CASE NO.</th>
<th>ACCESS NO.</th>
<th>DIAGNOSIS</th>
<th>CONTRIBUTOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>P 1</td>
<td>8888) 9061)</td>
<td>Benign proliferating epidermoid cyst.</td>
<td>Leo Kaplan, M.D.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Paul Thompson, M.D.</td>
</tr>
<tr>
<td>P 2</td>
<td>8185</td>
<td>Fibro-epithelial papilloma.</td>
<td>Dean Wiseley, M.D.</td>
</tr>
<tr>
<td>P 3</td>
<td>6561</td>
<td>Epidermoid ca., ear.</td>
<td>Peter Dykstra, M.D.</td>
</tr>
<tr>
<td>P 4</td>
<td>9241</td>
<td>Keratoacanthoma.</td>
<td>E.M. Butt, M.D.</td>
</tr>
<tr>
<td>P 5</td>
<td>9056</td>
<td>Intra-epithelial ca., vulva</td>
<td>Leo Kaplan, M.D.</td>
</tr>
<tr>
<td>P 6</td>
<td>8155</td>
<td>Extra-epithelial Paget's disease</td>
<td>H. Russell Fisher, M.D.</td>
</tr>
<tr>
<td>P 7</td>
<td>8969</td>
<td>Cylindrome.</td>
<td>T.S. Kimball, M.D.</td>
</tr>
<tr>
<td>P 8</td>
<td>8969</td>
<td>Hidradenoma.</td>
<td>Philip Flynn, M.D.</td>
</tr>
<tr>
<td>P 9</td>
<td>9075</td>
<td>Sweat gland adenoma</td>
<td>Dorothy Tatter, M.D.</td>
</tr>
<tr>
<td>P 10</td>
<td>9190</td>
<td>Tumor, Bartholin gland</td>
<td>Raymond Teplitz,</td>
</tr>
<tr>
<td>P 11</td>
<td>9098</td>
<td>Sweat gland ca.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>(take out)</td>
<td></td>
</tr>
<tr>
<td>P 12</td>
<td>9131</td>
<td>Sweat gland ca.</td>
<td>Lorin Spencer, M.D.</td>
</tr>
<tr>
<td>P 13</td>
<td>9070</td>
<td>Tricho-epithelioma, nipple</td>
<td>Harder, M.D.</td>
</tr>
<tr>
<td>P 14</td>
<td>8889</td>
<td>Malignant hidradenoma, papilliferum.</td>
<td>Eunice Waters, M.D.</td>
</tr>
<tr>
<td>P 15</td>
<td>9069</td>
<td>Basal cell ca.</td>
<td>H. Russell Fisher, M.D.</td>
</tr>
<tr>
<td>P 16</td>
<td>8088</td>
<td>Granular cell myoblastoma</td>
<td>Ruth McCammon, M.D.</td>
</tr>
<tr>
<td>P 17</td>
<td>8088</td>
<td>Reticulo-endothelial tumor</td>
<td>Alvin G. Foord, M.D.</td>
</tr>
<tr>
<td>CASE NO.</td>
<td>ACCESS NO.</td>
<td>DIAGNOSIS</td>
<td>CONTRIBUTOR</td>
</tr>
<tr>
<td>---------</td>
<td>------------</td>
<td>-----------------------------------</td>
<td>----------------------</td>
</tr>
<tr>
<td>18</td>
<td>9219</td>
<td>Active junctional nevus</td>
<td>Reuben Strauss, M.D.</td>
</tr>
<tr>
<td>19</td>
<td>9066</td>
<td>Lymphosarcoma skin and parotid</td>
<td>L. Frost, M.D.</td>
</tr>
<tr>
<td>20</td>
<td>8530</td>
<td>Mycosis fungoides</td>
<td>Donald Alsott, M.D.</td>
</tr>
<tr>
<td>21</td>
<td>9059</td>
<td>Dermatofibrosarcoma</td>
<td>Leo Kaplan, M.D.</td>
</tr>
<tr>
<td>22</td>
<td>9200</td>
<td>Letter-Sievers's disease</td>
<td>Herbert Lund, M.D.</td>
</tr>
<tr>
<td>23</td>
<td>9185</td>
<td>Urticaria pigmentosa</td>
<td>Ewald Lonser, M.D.</td>
</tr>
<tr>
<td>24</td>
<td>8639</td>
<td>Cellular blue nevus</td>
<td>Robert Blossom, M.D.</td>
</tr>
<tr>
<td>25</td>
<td>9182</td>
<td>Juvenile melanoma</td>
<td>Geo. J. Hummer, M.D.</td>
</tr>
</tbody>
</table>

LEFT OVER- CASES CUT.

- 9219 - Active junctional nevus  
  Reuben Strauss, M.D.

9218     Intradermal compound nevus  
  H. Russell Fisher, M.D.

8627     Pigmented spindloid ca.    
  L.A.C.H.
CASE NO. 1

ACCESSION NO. 9417
OUTSIDE NO. EDF 316-49

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Malignant melanoma of choroid
Spindle A (Callender classification).

HISTOLOGY: The cells in this tumor, which arises in the choroid, are for
the most part spindle-shaped, and contain densely packed chromatin material
of non-particulate appearance. Nucleoli are not apparent in this type of
cell. There are, however, many cells of the spindle B variety in which the
chromatin material is readily discernible, and multiple nucleoli easily seen.
Although this tumor exhibits both spindle B and spindle A cells, I feel that
the latter predominate. The serous lakes seen throughout the tumor are a
common feature of melanoma of the choroid, and to my knowledge have little
significance. In contrast to melanomas elsewhere, there is little mitotic
activity evidenced in this lesion. Note that the overlying pigment epithe-
lium is absent and that there is a serous separation of the adjacent retina.
The rest of the eye is essentially normal.

DISCUSSION: Malignant melanomas of the uveal tract have been classified
according to their cell type by Callender, Wilder, and Ash. The spindle A
is the least malignant, and the cell is characterized by a narrow, oval
nucleus containing densely packed chromatin material and an indistinquish-
able or ill-defined nucleolus. The spindle B cell has a slightly larger
nucleus in which particulate chromatin material and distinctly staining,
often multiple, nucleoli can be seen. The fascicular type, statistically
more malignant than the spindle types, is characterized by a palisaded
arrangement of spindle B cells. The most malignant of the choroidal melano-
mas is the epithelioid type, composed of large, rounded or polygonal cells
with considerable pleomorphism. Multinucleated forms are common. The cell
walls are well defined. Multiple nucleoli are seen. The chromatin materi-
al is particulate. The cells are generally less cohesive. The mixed cell
type usually refers to a mixture of epithelioid and spindle B and has been
said to be the second most malignant.

REFERENCES:

Ophthalmic Pathology Atlas - Saunders; Callender, G.R., Wilder, H.C.,
Ash, J.E.: Five hundred melanomas of the choroid and
ciliary body followed five years or longer. Am. J.
Ophth. 25:962-967, 1942.
Dr. Irvine's diagnosis: Malignant melanoma of choroid
Spindle B (Callender classification).
Separation of retina secondary to intraocular neoplasm.

HISTOLOGY: Of primary interest is the malignant melanoma apparently arising in the choroid and mushrooming through the lamina vitrea into the vitreous cavity. The overlying retina is thinned, atrophic, and in some places absent. There is a serous separation of the adjacent retina. The tumor itself is moderately vascular and is composed primarily of spindle B cells, some of which are arranged in fascicles. The anterior segment of the globe is normal. There is no evidence of extrabulbar extension in the sections examined.

DISCUSSION: It will be noted that this tumor is situated at the equator of the eye where the emissaria are few. It has been our impression that externalization of tumors in this area is less frequently seen than in instances of tumors occurring in the posterior segment, where the greater number of penetrating scleral emissaria are located.

REFERENCE:

Tumors of the Eye - Reese.
Dr. Irvine's diagnosis: Malignant melanoma of choroid
Mixed cell type (Callender Classification).

HISTOLOGY: The tumor fills the interior of the globe and is composed of two types of cells, spindle B and epithelioid. The spindle B cells show distinctly staining chromatin material and nucleoli, some of which are arranged in palisades. The epithelioid cells reveal much more pleomorphism, have a tendency to be less cohesive and have a well defined cytoplasm that is often acrophilic. Multiple nucleoli are seen, and mitotic figures are more apparent than in the pure spindle varieties of intraocular melanoma. There is extensive necrosis seen centrally and there is obvious extension of this tumor through the scleral emissaria. It is also of interest that the tumor fills the anterior chamber. There is a hypermature cataract that is of little significance to the pathologist.

DISCUSSION: Externalization of intraocular malignant melanoma has always been regarded as a poor prognostic sign, not because they tend to grow well or metastasize from the orbit, but because by this time, distant metastases have probably occurred. Indeed, ocular melanoma grow poorly in the orbit, and only infrequently is a recurrence noted in that location. We think it likely that extrabulbar extension of microscopic degree occurs much more frequently than suspected. Serial sections of several eyes containing small tumors showed evidence of extrabulbar extension in only a few sections.
Dr. Irvine's diagnosis: Malignant melanoma of choroid and ciliary
body Fascicular type, (Callender Classification).

HISTOLOGY: The central portion of the cornea has been removed for corneal
grafting. The malignant melanoma described in the gross examination is seen
to arise in the temporal choroid and pars plana of the ciliary body. The
overlying retina is thin and atrophic. The tumor, composed primarily of
heavily pigmented spindle B cells arranged in palisading fascicles, shows
considerable chronic inflammatory reaction, particularly around its blood
vessels, and in the adjacent and underlying choroid. There is little separa-
tion of the retina.

DISCUSSION: The fascicular grouping in the Callender classification is the
only one determined by the arrangement of cells, rather than by cell types.
This fascicular pattern is seen in many cases primarily belonging to the
other groups, and I question whether this group is sufficiently distinct to
warrant a separate classification. There have been numerous attempts to
correlate the cell type with the probable origin of choroidal melanoma. It
has been thought that the spindle and fascicular varieties are more closely
related to Schwannian elements, particularly the long posterior ciliary nerves,
and that perhaps the epithelioid cell melanomas may arise from the chromato-
phores of the choroid. However, both cell types are seen in the same neo-
plasm and often the portion of the tumor mushrooming within the eye will be
different from that within the choroid. The metastases may also differ from
the primary tumor. Moreover, benign heavily pigmented lesions of the chor-
oid, when bleached, closely resemble bleached choroidal chromatophores,
rather than the cells of the epithelioid type.

The intra-ocular malignant melanoma have been classified accord-
ing to their argyrophilic fiber content. The more malignant the tumor, the
less silver-staining reticulum it develops.

REFERENCE:

Tumors of the Eye - Reese; Wilder, H.C. An improved technic for silver
Dr. Irvine's diagnosis: Necrotic malignant melanoma of choroid, Separation of retina secondary to intra-ocular neoplasm, Endophthalmitis secondary to necrotic intra-ocular neoplasm.

HISTOLOGY: The posterior segment is filled with a necrotic, organized inflammatory mass that appears to arise in the choroid. The retina is completely separated by a primarily serous sub-retinal fluid on one side, and an organized (partial or complete) inflammatory exudate in the sub-retinal space on the other. This is the clue to the diagnosis. In some sections, sheets of poorly staining pigmented tumor cells can be seen within the sub-retinal inflammatory mass. The choroid is distended adjacent to the tumor, and is compressed beneath the serous retinal separation. The iris leaves are bowed anteriorly in many sections, occluding the trabeculae, and illustrating one of the mechanisms of secondary glaucoma in instances of intra-ocular neoplasm. There is considerable atrophy of the iris, particularly of its pigment epithelium. This is more evident on the side of the tumor. Indeed, the necrosis of the ciliary body and iris on the side of the neoplasm suggest that the long posterior ciliary artery and some of the branches of the anterior ciliaries have been destroyed. The lens is quite cataractous, as evidenced by a loss of its epithelium anteriorly, and the vacuolization of the cortex posteriorly.

DISCUSSION: The tumor cells are only found in approximately twenty-five of the slides. The diagnosis, however, should be suspected from the difference in the sub-retinal pathology on the two sides of the section. To me, this case illustrates the value of cutting one hundred and fifty sections and staining every tenth one, thus giving a composite picture of the pathology. Necrosis is considered to be a serious prognostic sign of intra-ocular melanoma. Fifty percent of necrotic melanoma have distant metastasis evident within five years. However, this is probably due to the size of the tumor and is a reflection of its rapid growth.

Intra-ocular inflammation commonly occurs secondary to intra-ocular malignant melanoma. Conversely, intra-ocular melanoma are not infrequently found in old pathisical, chronically inflamed eyes.
CASE NO. 6
ACCESSION NO. 9422
OUTSIDE NO. EDEF 193-54

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Disciform degeneration of the macula.

* * * * * * *

CASE NO. 6A
ACCESSION NO. 9422
OUTSIDE NO. EDEF 122-56

CONTRIBUTOR: A. R. Irvine, Jr., M.D.

Dr. Irvine's diagnosis: Early disciform degeneration of the macula
(macular serosanguineous separation).

* * * * * * *

CASE NO. 6B
ACCESSION NO. 9422
OUTSIDE NO. EDEF 105-49

CONTRIBUTOR: A. R. Irvine, Jr., M.D.

Dr. Irvine's diagnosis: Penetrating wound of eyeball.
Band keratopathy.
Adherent Leucoma.
Secondary glaucoma.
Progressive myopia.
Retinal hemorrhage.
Disciform degeneration of the macula.

* * * * * * *

CASE NO. 6C
ACCESSION NO. 9422
OUTSIDE NO. EDEF 523-49

CONTRIBUTOR: A. R. Irvine, Jr., M.D.

Dr. Irvine's diagnosis: Disciform hemorrhagic separation of pigment
epithelium of retina.

HISTOLOGY: This is a composite case, the various slides of which illustrate
different phases of disciform degeneration of the macula. In the earliest
cases there is a serosanguineous separation of the pigment epithelium with
breaks in the lamina vitrea. This elevates the retina and often gives the
clinical appearance of a heavily pigmented malignant melanoma. As long as
continued—
the pigment epithelium remains intact the vision may remain normal, although a change in refraction manifested by increasing hyperopia is caused by the abnormally elevated retina. With destruction of the pigment epithelium, and consequently the neurosensory epithelium, a central scotoma occurs. The blood becomes organized and forms a white, dense fibrous tissue plaque that is comparatively acellular. The pigment epithelium is always absent, although pigmented cells are seen within this white subretinal tissue. This picture is thought to be a result of organized hemorrhage, as well as metaplasia of the pigment epithelium.

DISCUSSION: Disciform degeneration of the macula, Kuhnt-Junius macular degeneration, and serosanguineous separation of the pigment epithelium, all refer to hemorrhagic extravasations and their sequelae in the macular region. The primary pathology may well be one of elastic tissue in that the elastic portion of the lamina vitrea stains basophilically, and that other breaks in the lamina vitrea are often seen accompanying this condition. These breaks are seen usually concentric to the disc and casually resemble blood vessels, therefore acquiring the name "angioid streaks". Systemic manifestation, such as gastric hemorrhage and pseudo-xanthoma elasticum are occasionally found in these patients. Disciform degeneration is most commonly confused with malignant melanoma of the choroid. It can be differentiated from that tumor by the absence of an adjacent serous separation of the retina, and usually there is evidence of blood around the periphery of the mass. Hemorrhage is rare in a melanoma of such small size. Statistically melanomas are rare in the macula, whereas disciform degeneration commands occurrence in that region.

REFERENCES:


CASE NO. 7

ACCESSION NO. 9423
OUTSIDE NO. EDEF 433-54

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Malignant melanoma of iris.

HISTOLOGY: A solid, heavily pigmented tumor almost fills the anterior chamber. The tumor is composed of spindle cells and epithelioid cells, some of which show a tendency to be arranged in packets resembling nevus formation. The tumor is quite vascular. In the site opposite the bulk of the tumor, there is extension of neoplasm into the trabeculae and into the iris meshwork.

DISCUSSION: The history suggests that this tumor arose on the anterior surface of the iris and had been present for approximately ten years. In many cases such tumors have been observed to arise from pre-existing small pigmented iris lesions. This represents the location of benign melanotic lesions, such as iris freckles and collections of iris chromatophores. Reese states that true nevi of the iris are not uncommon. He cites the increased incidence of nevi and freckles in eyes that harbor malignant melanoma. Malignant melanoma of the iris are more benign than those occurring in the posterior uvea and seem to occur in a younger age group. This is probably explained by the earlier detection and treatment of such tumors.
CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Malignant melanoma of the iris and ciliary body.

HISTOLOGY: The tumor is seen in the corona ciliaris and iris root on each side of the section. It has invaded the trabecular meshwork and is in close approximation to the vessels of Schlemm's canal. Tumor tissue extends along the posterior surface of the cornea about 2 mm, and is seen heavily infiltrating the anterior stroma and anterior surface of the iris and in some sections, as far as the pupillary margin. The tumor has not produced appreciable thickening of the ciliary body, but has involved the iris root. It represents the "ring sarcoma" described in the older literature. It is of clinical interest that glaucoma and heterochromia of the iris were the presenting signs.

DISCUSSION: Zimmerman and Rones have recently given their preliminary report regarding the malignant melanoma of the iris seen at the Armed Forces Institute of Pathology and collected from sources throughout the country. They divided these lesions into solitary solid tumors, such as we have seen in Case No. 7, and into the diffuse melanoma of which this case is an example. Frequently, glaucoma is the presenting sign and often glaucoma surgery is performed because the primary pathology is not suspected. Gonioscopy of all patients, plus greater clinical acumen in the presence of diffuse pigmentation and lack of iris atrophy in the affected eye should suggest the presence of a malignant melanoma.

REFERENCE:
Zimmerman and Rones - to be printed.
CASE NO. 9

ACCESSION NO. 9422
OUTSIDE NO. EDF 257-50 and 5-57

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Multiple malignant melanomata of bulbar conjunctiva.

HISTOLOGY AND DISCUSSION: Microscopically this tumor resembles the others removed from this patient. The cells are anaplastic and pleomorphic, and vary in their intensity of staining reaction. They assume spindle, oval and epithelioid configuration, multiple nucleoli of varied size and abnormal bizarre chromatin patterns. Abnormal mitoses are seen. There is a fine reticular network. The earlier lesions showed a small amount of dark brown intracellular pigment, whereas the more recent tumors were almost amelanotic, although some pigment can be seen in a few cells in all sections I examined. Most of the sections present some areas of chronic inflammatory reaction. Necrosis was minimal, except in two of the tumors in which there was accompanying hemorrhagic extravasation.
CASE NO. 10

ACCESSION NO. 9425
OUTSIDE NO. MREF 257-50 and 5-57

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Precancerous and cancerous melanosis.

HISTOLOGY: These sections are from the upper nasal lid margin, an area that was never clinically abnormal. The epithelium of the conjunctiva shows a loss of cellular polarity, clear spaces surrounding nuclei, and a pleomorphism that is associated with precancerous melanosis. In places the basement membrane is indistinct and shows the junctional activity and destruction of epithelium seen in cancerous melanosis. There are no mature nevus cells in the dermis. It appears that cells of malignant melanoma are being formed within the basal layers of the epithelium.

DISCUSSION: All but one of the neoplasms in this case appear to arise in and beneath the conjunctiva, suggesting multiple origins rather than extension from a primary focus. This is supported by the histopathology of the upper nasal lid margin which shows the changes seen in precancerous and cancerous melanosis, except for the lack of pigment. Precancerous and cancerous melanosis are usually regarded as diffuse, flat, pigmented areas in the palpebral and bulbar conjunctiva. Once the malignant changes occur, this type of lesion is thought to be highly malignant. Exenteration of the orbit is the accepted form of treatment. The course of this particular case was of interest to us because diffuse premalignant changes appeared to give rise to solitary tumors that, although appearing to be extremely malignant histologically, were clinically quite benign. This case suggests that highly malignant appearing lesions may be slow growing and remain confined to the epibulbar tissues without undergoing early distant metastasis.

REFERENCE:

CASE NO. 11

ACCESSION NO. 9551
OUTSIDE NO. EDEF 369-52

CONTRIBUTOR: A. R. Irvine, Jr., M.D.
Estelle Doheny Eye Foundation
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Carcinoma of choroid, metastatic from prostate.

HISTOLOGY: The anterior segment is not remarkable. There are two metastatic foci in the choroid, one at the equator nasally, and another behind the equator temporally. Several short posterior ciliary arteries contain tumor emboli. The neoplastic cells are large, oval and epithelial in type. Mitotic figures are occasionally seen. There is a small amount of blood in the inner layer of the retina. The optic nerve is edematous.

DISCUSSION: Both the acid and alkaline phosphatase were increased, findings compatible with carcinoma of the prostate metastatic to the bone. The choroid metastatic foci were isolated from each other, suggesting that the tumor cells reached the tissue through several short posterior ciliary arteries.

To our knowledge, nine cases of metastatic carcinoma from the prostate have been reported, six of these were confirmed histologically.

REFERENCE:

CASE NO. 12, 12A, 12B, 12C

ACCESSION NO. 9478

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Compound nevus of bulbar conjunctiva.

HISTOLOGY AND DISCUSSION: All of these nevi show the characteristic groups of nevus cells within the substantia propria and the epithelium. There is some evidence of junction activity. The accumulations of mucous secreting epithelium and cysts is a common finding in conjunctival nevi. It appears that there is a nevoid dysplasia of various types of epithelium in an aberrant location. However, it is more likely that the cysts represent surface epithelium that has been surrounded by nevus cells and then carried into the stroma as these cells matured and extended into the deeper tissues. There are examples of solid masses of epithelial cells undergoing various forms of cystic degeneration, and the walls of some of these cysts are occasionally lined by nevus cells. Mucous secreting epithelium is always present in any type of conjunctival irritation, and it is not surprising to see mucous secreting epithelium surrounded by nevus cells carried into the stroma in the manner described above.
CASE NO. 13

ACCESSION NO. 9427
OUTSIDE NO. EDEF 512-56

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Retinoblastoma with extrabulbar extension.

HISTOLOGY: This tumor shows the usual characteristics of retinoblastoma, including necrosis, calcification, pseudorosettes, true rosettes and seedlings of tumor about the interior of the eye and apart from the primary neoplasm. It is unusual in that it extends extrabulbarly through the posterior scleral emissaria and not through the optic nerve. The choroidal extension indicates a grave prognosis.

DISCUSSION: In our experience most retinoblastomas have both true and pseudorosettes and arise from both inner and outer nuclear layers. They often grow into the vitreous cavity, become fragmented and produce vitreous opacities. In these cases tumor cells may be seen in the anterior chamber. At times the growth extends toward the choroid and the vitreous remains clear. The most common mode of extension is via the optic nerve and for this reason cross sections of the nerve should be made. The tumor produces calcium that can frequently be seen by X-ray. The optic foramen may be enlarged. The absence of pupillary reaction probably indicates optic nerve involvement. Twenty-five percent are bilateral.

REFERENCE:
CASE NO. 14

ACCESSION NO. 604
OUTSIDE NO. EDEF 230-49

CONTRIBUTOR: A. R. Irvine, Jr., M.D., Estelle Doheny Eye Foundation, 272 South Lake Street, Los Angeles 57, California.

Dr. Irvine's diagnosis: Diktyoma.

HISTOLOGY: The tumor arises from the ciliary epithelium on one side. It measures about 3 mm. antero-posteriorly and about 3 mm. equatorially. It is composed of cells resembling the non-pigmented ciliary epithelium. They are arranged in oval and round tubules that vary considerably in design and size. Elsewhere, they occur in bands and sheets that form intricate varying patterns. Some structures resembling true rosettes are seen. There is some necrosis as evidenced by free blood in the masses of inflammatory cells. There is no pigmentation. This tumor mass primarily involves the corona ciliaris. It extends along the peripheral portion of the ciliary irides. It fills the circumlental space on one side and is accompanied by a fibrotic membrane that appears attached to the equatorial portions of the lens. There is no other extension of the tumor. There are also broad peripheral anterior synechiae and at the false angle on the side of the tumor, the endothelium and Descemet's membrane appear to split and extend on the anterior surface of the iris. The iris stroma contains more than the usual number of cells. The crypts are not well formed. The posterior segment of the eye is not remarkable. There is no evidence of nerve atrophy or ganglion cell damage.

DISCUSSION: Such tumors are exceedingly rare. They are only locally malignant. There is no evidence of extrabulbar extension of this tumor.

REFERENCES:


CASE NO. 14A
ACCESSION NO. 604
OUTSIDE NO. 947

CONTRIBUTOR: Leonard Christensen, M.D.,
University of Oregon Medical School,
Portland 1, Oregon.

Dr. Christensen's diagnosis: Diktyoma ?.
Aberrant type of retinoblastoma ?

HISTOPATHOLOGY: Microscopic sections of this moderately degenerated glaucomatous infant eye show complete detachment of the retina and a subretinal epithelial tumor of indefinite origin. The tumor is composed of neuro-epithelial cells and is partitioned by dense strands of mesodermal tissue. Immediately adjacent to the mesodermal strands, the epithelium is partially pigmented and arranged in pseudosockets. Away from the mesoderm the character of the cell and its growth pattern change. The cells are less columnar and form many loculated epithelial lined cysts which are nonpigmented. On one side the tumor is continuous with the nerve fiber layers of the retina and is partially enveloped by retinal tissue. The tumor has also invaded the choroid and has assumed an abortive hour-glass shape; i.e., the base of the tumor in the choroid is much wider than its neck where it extends through Bruch's membrane in some areas but there are no indications of independent satellite lesions.

In addition to the tumor there are extensive anterior peripheral synechiae with occlusion of the chamber angle on both sides. There is a retrolental fibrous connective tissue mass which partially covers the posterior lens surface. The retina is completely detached but the presence of ganglion cells indicates that no prolonged increased intra-ocular pressure had occurred. The optic nerve shows excellent columnar arrangement of its nerve fibers and was viable.

DISCUSSION: Although the origin of this tumor is undetermined, there are several features that link its origin with either the retina or the nonpigmented ciliary epithelium of the pars plana. The tumor is in direct continuity with the nerve fiber layers of the retina and the retina overlying the tumor appears to be invaginated since the nerve fiber layer is adjacent to the tumor. In addition, there are two types of neuro-epithelial cells in the tumor. Trichrome stains using bleached and unbleached sections indicate that the nuclei of one element stain a deep red, whereas the other cells have blue staining nuclei. The size, appearance and shape of the cells vary depending on their location in the tumor. Trichrome stains of normal retina and of retinoblastoma demonstrate deep red stain in the nuclei of the outer nuclear layer of the retina and a relatively light blue stain of the inner nuclear layer. Phosphotungstic acid stains on both the bleached and unbleached sections show a peculiar granular deposit within the nuclei of tumor cells. Similar stains of globes with retinoblastoma present a similar type of cell. This granular deposit, which is probably an artefact, is also present in the nonpigmented ciliary epithelium along pars plana. Another possible origin of this tumor is the nonpigmented ciliary epithelium along pars plana. The evidence for this is the similarity in staining reaction of the nuclei of this epithelium to that of the tumor with trichrome stains of bleached sections. The nonpigmented ciliary epithelium along pars plana also presents the granular deposition within the nuclei to phosphotungstic acid stains as the pigmented tumor cells. Finally, on one side of the tumor there is pigmented epithelial tissue extending along its surface which simulates the morphology of ciliary epithelium. Bleached sections of the pigmented element of the tumor show no similarity in staining characteristics to the pigmented epithelium along Bruch's membrane or the iris.
Dr. Irvine's diagnosis: Persistence and hyperplasia of the primary vitreous.
Cataract secondary to the above.
Anterior subluxation of lens secondary to pupillary block.

HISTOLOGY: The cornea is thinned. The lens is subluxated into the anterior chamber; its capsule appears to be ruptured and its cortex is cataractous. There are broad peripheral anterior synechiae of the type formed by iris bombe'. The endothelium and Descemet's membrane extend onto the anterior surface of the iris on one side. A proliferating endothelium, along with a clear membrane, extends beyond the pupillary border toward the lens. The iris and ciliary body are atrophic.

The anterior segment is somewhat staphylomatous. Posteriorly, ganglion cells are reduced in number. There is some atrophy of the optic nerve and it is abnormally cupped. The ciliary body is extremely small. The corona and pars plana are poorly differentiated and the ciliary processes appear to extend across the equator of the eye as a thin pigmented tissue containing some non-pigmented epithelium. Centrally and immediately posterior, there is an ill-defined mass of glial tissue in which ciliary pigmented and non-pigmented epithelium can be seen. On one side the retina reflects onto this cyclitic membrane. In some sections one sees a small group of epithelial cells, resembling lens epithelium, just behind the cyclitic membrane. In some sections posteriorly, the retinal tissue appears to be elevated on the nasal side immediately adjacent to the disc, and a vessel is seen entering this elevation. This is suggestive of remnants of the hyaline circulation. Retinal tissue appears to be pulled into the optic cup on the temporal side.

DISCUSSION: Persistence and hyperplasia of the primary vitreous is usually unilateral, occurs in eyes that are either normal in size or slightly small and is detected shortly after birth because of the white pupillary reflex. It is often mis-diagnosed as retinoblastoma, but differs in that the anterior chamber is always shallow, the posterior surface of the lens may show an anterior concavity, the opacity is densest in the central portion, long ciliary processes are seen in the periphery and remnants of the hyaloid artery, always seen microscopically, may be seen clinically. The remnants of the hyaloid vascular system are often firmly attached to the posterior lens capsule which at times ruptures, resulting in a localized cataractous change.

In this case, the whitish mass behind the lens seen clinically is undoubtedly the remnants of the hyaloid system. These remnants often appear on one side and attach to the pars plana of the ciliary body, producing a cyclitic membrane. This membrane pulls the ciliary processes toward the lens and results in a decreased tension of the zonular fibers, permitting the iris and lens diaphragm to be displaced anteriorly, thereby occluding the chamber angle and causing glaucoma.

continued-
Case No. 15 - continued.

Persistence and hyperplasia of the primary vitreous enters into the differential diagnosis of retinoblastoma. Other conditions comprising the group of so-called pseudoglioma and clinically confused with retinoblastoma, are retrolental fibroplasia, metastatic retinitis, Coats' disease, massive retinal fibrosis in children and nematode endophthalmitis.

REFERENCES:


Case No. 16

ACCESSION NO. 9429
OUTSIDE NO. EDEF 268-50

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Exudating hemorrhagic retinopathy.

HISTOLOGY: The retina is separated, gliosed, folded and pulled anteriorly. Cholesterin clefts, giant cells, blood pigment and phagocytes are seen in its outer layers, particularly near the equator where the retina is pressed to the underlying choroid by a fibrous connective tissue. In some sections the retina is split, suggesting the occurrence of a massive retinal hemorrhage. In some places the pigment epithelium has proliferated, whereas in others it has disappeared. There is some basophilic staining of the lamina vitrea, presumably because of calcium deposition.

The subretinal fluid is filled with a serosanguineous exudate that contains large numbers of pigment laden phagocytes.

The choroid is compressed because of formalin fixation. A few foot of chronic inflammation are seen in the choroid. The ciliary body is somewhat flattened. Iris processes extending to the trabeculae are reminiscent of the fetal type of chamber angle meshwork.

DISCUSSION: The retinal separation is secondary to hemorrhage. The pathology is similar to exudative hemorrhagic retinopathy externa occurring in the adult, and originally described by Coats.

In his description of massive exudative retinitis, Coats included angiomatosis retinae and lesions caused by retinal hemorrhage at birth (massive retinal fibrosis). The former has since been classified as a separate entity. I have been unable to differentiate between massive retinal fibrosis of the newborn and exudative retinopathy of the young adult, except that in the former the retina is not separated. The retina is elevated due to a yellowish subretinal mass and there is evidence of hemorrhage, or its sequela. The lesion in the adult often present vascular dilations, tortuosities and abnormal anastomoses. Hemosiderin cholesterol crystals, inflammatory cells and fibrous masses are noted. The last may become calcified or undergo deposition of bone. Masses of phagocytes are seen in the hemorrhagic exudates. Thickening of vessel walls and vascular occlusions may be present. The pathogenesis of Coats' disease is speculative.

Those who differentiate Coats' disease from massive retinal fibrosis in the newborn feel that there is an underlying vascular anomaly in the former, whereas the latter represents proliferative changes occurring secondary to retinal hemorrhages in an eye without congenital anomaly. Retinal hemorrhages are not uncommon in the newborn and intra-cranial hemorrhage has been estimated in ten percent of infants. In some cases the retina does not detach, but becomes firmly adherent to the underlying choroid because of the formation
Case No. 16 - continued.

of fibrous tissue. The contracture of this fibrous tissue resulting from organization of blood may give the clinical appearance of a progressive lesion, thus causing the fundus picture to closely resemble that seen in retinoblastoma.

REFERENCES:


CASE NO. 17

ACCESSION NO. 9430
OUTSIDE NO. BIEF 331-49

CONTRIBUTOR: A. R. Irvine, Jr., M.D.
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Papillary squamous cell carcinoma of the limbus.

HISTOLOGY: The sections described pass horizontally and inferiorly through the largest portion of the new growth. About 7 mm. posterior to the temporal limbus, the bulbar conjunctiva reflects backward upon itself and gradually thickens to form the temporal portion of the new growth. The tumor is papillary, each projection having its own connective tissue core. The prickle cells are dyskeratotic and vary somewhat in size and shape and in the density of their staining reaction. They have lost their normal polarity and arrangement. Single and multiple nucleoli are evident and frequently small vacuoles are seen within the nucleus. Mitotic figures are frequently seen and in some places mitotic division is represented by tumor giant cells. There is a tendency for the cells to assume a concentric arrangement resembling abortive epithelial pearls. Eosinophilic staining of the outermost layers is evidence of keratinization. However, the outermost layers contain cell nuclei. Large numbers of lymphocytes and plasma cells are seen within the connective tissue stroma. The tumor has not invaded the cornea and is distinctly separated from Bowman's membrane by connective tissue containing chronic inflammatory cells. This, I believe, represents connective tissue developed by the tumor rather than a degenerative pannus. The neoplasm is moderately vascular and shows little necrosis. The corneal stroma, endothelium and Descemet's membrane are not involved, although there is some vascularization and fibrosis of the stroma near the limbus on one side. No pathology is seen within the globe.

DISCUSSION: Squamous cell epithelioma usually arise at the limbus and tend to extend over the cornea. They rarely invade the corneal stroma and histologically appear more benign than similar tumors arising elsewhere. It has been our impression that papillary epithelioma of this region do not show the extensive dyskeratotic changes exhibited by the more flat diffuse epithelioma arising from Bowen's dyskeratosis or leukoplakia. Epidermalization of adjacent epithelium is seen in papillomas as frequently as it is in carcinomas and therefore does not help us to differentiate between potentially benign and malignant lesions.

REFERENCES:

Dr. Irvine's diagnosis: Bowen's dyskeratosis of the bulbar conjunctiva and limbus.

HISTOLOGY: In these three cases the histology is essentially the same. Of primary interest is the epithelium of the bulbar conjunctiva. It is normal toward the outer canthus, but approaching the limbus it increases in thickness four to five times. These cells have lost their normal polarity, are somewhat pleomorphic and there is keratinization of individual cells. An occasional mitotic figure is seen. The basement membrane is intact and there is no indication of invasiveness. There is considerable chronic inflammatory reaction in the stroma.

DISCUSSION: Bowen's dyskeratosis, intra-epithelial epithelioma and squamous cell epithelioma occur at the limbus. Basal cell epithelioma does not occur in this area. Pterygia will often be covered by hyperplastic and slightly dyskeratotic epithelium. However, squamous cell epithelioma only rarely develops from such lesions. Bowen's dyskeratosis will often give rise to squamous cell epithelioma. The process remains limited to the superficial tissue, Bowman's membrane and the sclera apparently affording strong barriers against invasiveness. Removal locally by splitting the cornea and the sclera and checking the margins by frozen sections is the treatment of my choice. Rather extensive surgery can be done and the defect adequately filled by conjunctival transplant from the upper fornix of the opposite eye. In my opinion, irradiation is not indicated because of danger to the lens, and also because in case of recurrence, dyskeratotic changes may be produced by radiation as well as the neoplasm.

REFERENCE:
CASE NO. 19

ACCESSION NO. 9432
OUTSIDE NO. EDEF 165-51

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Leiomyoma of iris.

*** *** ***

CASE NO. 19A

ACCESSION NO. 9432
OUTSIDE NO. 10880

CONTRIBUTOR: James E. Kahler, M.D.,
St. Vincent's Hospital,
2131 West Third Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Leiomyoma of iris.

*** *** ***

CASE NO. 19B

ACCESSION NO. 9432
OUTSIDE NO. EDEF 368-55

CONTRIBUTOR: Ralph H. Fuller, M.D.,
St. Mary's Hospital,
Tucson, Arizona.

Dr. Irvine's diagnosis: Probable malignant melanoma of iris.

HISTOLOGY: Of these three cases, only 19A fulfills all the criteria for leiomyoma. The cells appear to arise from the dilator muscle of the iris. Myoglia fibrils are present and their nature proved by differential stains. The cells are spindle-shaped and their nuclei are rod-shaped with rounded ends. Mitotic figures are not seen.

In Case No. 19, there is a suggestion of muscle striations, the nuclei resemble those of leiomyoma, but the differential stains do not prove the presence of myoglia fibrils.

In Case No. 19B, the nuclear pattern resembles that seen in leiomyoma. However, the presence of brown pigment within the tumor cells is probably sufficient criteria to classify the lesion as a malignant melanoma.

DISCUSSION: Smooth muscle, from which a myoma might arise in the eye, is found in the ciliary body, iris, orbit and media of blood vessels. The musculature of the iris is of neuro-ectodermal origin. Myomas may arise from continued-
Case No. 19, 19A, 19B - continued.

places where smooth muscle is not generally found. This is true in Verhoeff's case of leiomyoma of the iris in which the tumor apparently arose from the anterior surface of the iris and did not connect with the musculature of the pigment epithelium. Other cases of leiomyoma of the iris show a continuity between the tumor and the muscle fibers.

In 1943, Kahler et al reported the seventh case of leiomyoma of the iris that fulfilled the criteria for establishing a definite diagnosis. These criteria are: 1. Spindle-shaped cells with long, centrally placed, rod-shaped nuclei; 2. A granular eosinophilic cytoplasm of the cells; 3. Myofibrils within the cytoplasm of the tumor cells as demonstrated with Mallory's phosphotungstic acid hematoxylin stain, van Gieson's specific staining reactions of smooth muscle tissue, or Mallory's aniline blue stain.

There is a tendency for the cells to be arranged in palisades. Mitotic figures are not seen. The tumors are benign, but may become malignant after repeated attempts at excision. In Terry's case of leiomyoma of the orbit, the first specimen reveals the necessary criteria for positive diagnosis, whereas after several recurrences it loses semblance to a leiomyoma and adopts the appearance of a sarcoma.

REFERENCES:


Dr. Irvine's diagnosis: Glioma of optic nerve.

HISTOLOGY: The anterior segment is not remarkable.

The nerve head is edematous and extends about one millimeter into the vitreous cavity. The retina is displaced laterally. The optic nerve is somewhat atrophic. A few ganglion cells are seen, but the nerve fiber layer and the ganglion cell layer are greatly thinned. The cells of the inner nuclear layer appear to be normal. At the posterior entrance to the optic nerve canal, the nerve disappears and the vaginal coverings collapse and come together. About one millimeter posterior to this region they separate and reveal elements of the glioma. This tumor appears to be exceedingly necrotic. It is severed at its posterior portion, but tumor cells with spindle and oval-shaped nuclei are exhibited.

DISCUSSION: The optic nerve is comparable to a central nervous tract, except in that it is subdivided into bundles by vascular septa from the pia. The nerve fibers are found separated by a glial network composed of astrocytes, oligodendrocytes and microglia. The first and the last apparently give rise to glioma.

Glioma of the optic nerve produce a slowly progressing, unilateral exophthalmus and optic atrophy. Seventy-five percent become manifest in the first decade of life. The tumor is occasionally bilateral, either because of direct extension from one nerve through the chiasm to the other, or because of multiple origin. X-ray may show an enlargement of the optic foramen. Meningioma and hemangioma are considered in the differential diagnoses. Meningioma usually occurs after the third decade and reduced vision is a late manifestation. Because of involvement of the nerve proper in glioma, the vision is reduced early.

REFERENCES:

Dr. Irvine's diagnosis: Lymphocytic lymphoma of lacrimal gland.

**HISTOLOGY:** The original biopsy of this tumor was from the anterior portion of the lacrimal gland. It revealed essentially normal glandular elements invaded by lymphocytes.

This specimen shows minimal glandular tissue, the rest having been replaced by masses of mature lymphocytes. Mitoses are common. Besides lymphocytes, there are larger and more lightly staining cells that have a moderate amount of faintly eosinophilic cytoplasm. These are thought to represent immature lymphocytes rather than reticulum cells.

**DISCUSSION:** The pathology in the lacrimal gland, the thyroid and in the excised supraclavicular lymph node is consistent with lymphomatous disease. McGavic found that in twenty-one verified cases of ocular lymphoma that in seventeen there was a primary tumor in the region of the eye, while in four, ocular involvement occurred during the course of generalized lymphomatous disease. In five patients the lesions were in the subconjunctiva. Seven of the lesions were in the lacrimal gland, five of these were primary.

In Reese's examples of lymphomatous disease, eighteen out of sixty-one were classified as lymphocytic cell lymphosarcoma, fifteen of these were of the lids and lacrimal gland. The average age of the patients was fifty-five years. Lymphoma of the lacrimal gland tends to present on the conjunctival surface, whereas mixed tumors usually present beneath the skin. The globe is usually displaced inferiorly and nasally.

Lymphomatous disease of the eye may occur years before generalized lymphoma develops. Rarely, lymphoma occurs within the eye, there being approximately seven cases in the literature. Catlin stresses the favorable prognosis of lymphosarcoma in the head and neck, compared to the generalized form of the disease. Stout found that treated patients (irradiation and surgery) show a much longer survival than untreated ones. The chances of survival are greatest if the patient has passed the fortieth year. McGavic stresses that lymphomas of the orbit are discovered early and therefore treated while the condition is still localized. In seventeen of his cases there was not a single local recurrence after treatment by X-ray. The disease appeared elsewhere in six of these patients.

**REFERENCES:**


Dr. Irvine's diagnosis: Pseudotumor of the orbit.

HISTOLOGY AND DISCUSSION: Chronic inflammation of the orbit is common and is most important in the differential diagnosis of neoplasm. In most cases the etiologic agent is not apparent. There is usually a localized orbital mass which enlarges rapidly, causing marked exophthalmus. Early involvement of the muscles is evidenced by diplopia. There is little or no clinical evidence of inflammation and therefore the process simulates a neoplasm and surgical removal seems indicated.

Histologically, these lesions show chronic inflammation and scar tissue. Most of the tumor is fibrotic. Foci of lymphocytes are seen throughout the fibrous reaction. Blood vessels are plentiful and often show intimal proliferation, sclerosis and hyaline degeneration. Reese divides pseudotumors into four categories: 1. Those in which lymph follicle formation is prominent. 2. Those showing muscle involvement - the orbital myositis group. 3. Those characterized by diffuse chronic inflammation with little tissue destruction and little scarring, and 4. Those in which there is essentially a fibrous scar with little active inflammation. The case presented here would seem to fall into group 4. In some instances there is predominantly a periarterial or perivenous inflammatory reaction resembling that seen in periarteritis or lupus erythematosus. There is a possibility that these lesions are related to the collagen diseases.

REFERENCE:

CASE NO. 23
ACCESSION NO. 9436
OUTSIDE NO. EDF 527-54

CONTRIBUTOR: A. R. Irvine, Jr., M.D.,
Estelle Doheny Eye Foundation,
272 South Lake Street,
Los Angeles 57, California.

Dr. Irvine's diagnosis: Benign neurofibroma of choroid,
Separation of retina, secondary to neurofibroma
of choroid.

HISTOLOGY: The corneal epithelium is absent over most of the section. The
corneal stroma shows a decrease in the number of corpuscles. Descemet's mem-
brane is slightly thickened. A few polymorphonuclear leukocytes are seen
scattered diffusely through the anterior stroma in some sections. The trabecu-
lae are not remarkable. A hemorrhagic eosinophilic coagulum is seen in the
anterior chamber. The lens capsule is intact. The epithelium is largely ab-
sent anteriorly. The cortex and nucleus were lost in sectioning. In the upper
part of this section, the retina is separated and there is an eosinophilic
coaagulum with granular debris, possibly representing lysed red blood cells,
in the subretinal fluid. There are extensive degenerative changes in the re-
tina that are thought to be due to delayed and improper fixation. There is
considerable cystic degeneration of the peripheral retina, some of which may
represent artefact. Below, similar degenerative changes are seen in the retina.
In addition, however, there are pigmented adhesions between the choroid and
retina which represent atrophic scarring, probably resulting from previous
diathermy.

Of primary interest is a large tumor within the choroid adjacent to and par-
tially overlapping the optic nerve. The overlying retina is atrophic and dis-
organized so that the normal arrangement of its layers is not apparent. A
small serous or serosanguineous separation of the retina is seen just peripheral
to the tumor. The new growth itself seems to be composed of sheets and whorls
of rather densely packed spindle cells. The nuclei, for the most part, do not
stain densely and the chromatin material is not particulate in appearance. Occa-
sional isolated, large, irregular, more densely staining cells are seen. The
stroma is fibrillar and collagenous in appearance. At times the nuclei appear
to be arranged in palisades. Verocay bodies, however, are not seen. In
places the collagenous stroma appears to be loose, however, there are no areas
of typical cystic degeneration such as might be expected in the Antoni type B
of neurilemmoma. The optic nerve appears to be essentially normal. In some
sections the emissaria of the short posterior ciliary nerves appear to be
widened and to contain a nerve showing increased cellularity.

While the tumor is presumed to have originated from the ciliary nerves, there
is no evidence to support this other than a widening of the posterior emissaria
resulting from thickening of the nerve sheath.

DISCUSSION: Any part of the eye or adnexa may show manifestations of neuro-
fibroma. When the uvea participates in the disease, there is usually a diffuse
involvement of the choroid, ciliary body and iris. This case is somewhat un-
usual in that there is a large tumor beneath the retina. There appears to be
a positive correlation between the occurrence of neurofibroma and melanotic

continued-
Case No. 23 - continued.

Lesions of the eye. Neurofibromatosis, with melanosis of the uveal tract and melanomas of the iris, has been described. Malignant melanoma of the choroid has been reported in a patient with neurofibromatosis and a neurinoma in the other orbit. Usually the patient develops glaucoma and an enlarged eye. Gliomas of the optic nerve may occur in conjunction with neurofibromatosis.

REFERENCES:

Reese, A. B.: Tumors of the eye.
Dr. Irvine’s diagnosis: Surgical wound of anterior eyeball.  
Extracapsular cataract extraction.  
Epithelization of anterior chamber.  
Uveitis secondary to epithelization of anterior chamber.  
Glaucoma secondary to epithelization of anterior chamber.

HISTOLOGY: The cornea is irregular in thickness and is edematous and diffusely infiltrated with scar tissue. Stratified squamous epithelium representing an epithelial downgrowth is seen on the posterior surface of the cornea and on the anterior surface of the iris. There is epithelium bridging the pupillary space, occluding and secluding it. Cataractous lens remnants are seen behind the pupillary membrane. Small peripheral anterior synchiae are present in some sections. There is a chronic inflammatory reaction in the ciliary body. There is some sclerosis of retinal vessels. Ganglion cells are present but reduced in concentration. The optic nerve is partially atrophic.

DISCUSSION: Conjunctival epithelium grows within the eyeball in two forms; first, as a large cyst that may increase in size to fill the anterior chamber; second, as a downgrowth of epithelium extending over the iris and posterior cornea. Poor wound closure resulting in delayed formation of the anterior chamber and hypotony are most common causes of these conditions. Often inclusions of capsular lens remnants within the wound are seen. This condition is usually undiagnosed clinically and the eyes are usually removed because of intractible glaucoma.

REFERENCES:


CASE NO. 25

ACCESSION NO. 9395
OUTSIDE NO. GSH 57-1468

CONTRIBUTOR: Albert F. Brown, M.D.,
Glendale Sanitarium and Hospital,
1509 East Wilson Avenue,
Glendale 6, California.

Dr. Irvine's diagnosis: Muco-epidermoid of inner canthus probably arising
from accessory lacrimal gland.

HISTOLOGY: The following is Dr. Brown's description of the microscopic
findings:

The indurated tissue from the lower anterior medial angle shows wide infiltra-
tion by malignant epithelial tumor exhibiting a wide range of morphology, in-
cluding acanthotic epidermoid islands, irregular slits or duct-like cavities
lined by mixed epidermoid and mucous tumor cells, acinar structures composed
of irregular cuboidal or goblet-shaped mucous cells and complex strips or
 glandular arrangements of tall columnar mucous epithelium. The mucous-acinar
phase extensively invades spongy bone in close relation to a turbinate and
also extends through the mucosa of the turbinate.

Sections of conjunctival and ocular tissues on the side next to the tumor area
show no recognizable involvement. Section of the eyelid including the lateral
margin of excision shows no tumor. The lacrimal gland shows no significant
lesion.

DISCUSSION: Stewart, Foote and Becker have described this tumor which occurs
in the salivary glands and have termed it muco-epidermoid. Previously it was
erroneously grouped with the mixed tumors. It comprises about five percent of
the tumors of salivary glands and arises from salivary duct epithelium. The
tumors all produce mucin that stains with mucicarmine. In addition, the cells
have epidermoid qualities, either resembling basal cell carcinoma or squamous
cell epithelioma.

The cases have been divided into benign and malignant categories. The former
occurred in patients of forty years or younger, whereas most of the malignant
tumors occurred in older patients. The patients with histologically benign
tumors were alive and free from disease. The prognosis is poor for the malig-
nant type. At the time of their report, seven patients in this category were
dead. Out of twelve living patients, two were dying and a third was known to
have metastases.

REFERENCES:


Stewart, Foote and Becker,: Muco-epidermoid tumors of salivary glands. Annals