

*Surgical Management of Intraocular Tumors Surgical Management of Intraocular Tumors Surgical management of intraocular tumors has been mentioned in several parts of this book, but specific steps have not been illustrated.*

Multimedia Enucleation and evisceration are the 2 possible surgical management options for a disfigured or a painful blind eye. Several anecdotal studies of unsuspected tumors found after evisceration exist. Eagle et al 13 recently described a series of 7 additional cases of unsuspected uveal melanoma diagnosed in eviscerated specimens and have emphasized the role of detailed a medical history, clinical evaluation, and appropriate imaging before performing evisceration in painful blind eyes with opaque media. Our series comprises 6 cases of unsuspected intraocular tumors diagnosed following evisceration and elaborates on the lessons learned from this experience. Methods This is a retrospective, nonrandomized, clinicopathological case series of patients with previously unsuspected intraocular tumors diagnosed by histopathology of eviscerated blind eyes. The cases were collected by searching the indexed ocular pathology registry from January to December at a tertiary care center in southern India. The medical records were reviewed for patient age, sex, symptoms, initial clinical findings, initial clinical diagnosis, imaging, prior treatment, indication for evisceration, intraoperative findings, histopathology, postevisceration management, and final outcome for local tumor recurrence and systemic metastasis. Results We identified 6 patients with unsuspected intraocular tumors who had undergone evisceration Table. The median patient age was 18 years range, years; mean [SD], Preoperative ultrasound B-scan was available for 3 patients; a review of the documented images showed no obvious intraocular mass in 2 patients. A preoperative computed tomographic scan in one patient did not reveal an intraocular mass. Indications for evisceration included a painful blind eye in 4 patients, cosmetic concern in 1 patient, and a perforated hypotonus eye with uveal prolapse in 1 patient. No surgeons had recorded unusual features of intraocular contents observed during evisceration. Eviscerated tissue was not submitted for histopathology in 2 patients. Histopathology of the eviscerated tissue or biopsy from the recurrent orbital tumor revealed retinoblastoma in 2 patients, and 1 each of uveal melanoma, adenocarcinoma of the ciliary body, choroidal ganglioneuroma, and conjunctival squamous cell carcinoma with intraocular invasion. Orbital exenteration was eventually required to treat 4 of these patients. Two patients with retinoblastoma were treated with high-dose chemotherapy, orbital exenteration, and external beam radiotherapy. The patient with uveal melanoma had orbital exenteration and external beam radiotherapy. Adenocarcinoma of the ciliary body was managed with enucleation and external beam radiotherapy. The patient with benign choroidal ganglioneuroma was observed. The patient with intraocular invasion of conjunctival squamous cell carcinoma had orbital exenteration. All of the patients were free of local recurrence or systemic metastasis at a median follow-up of 28 months range, months; mean [SD], There was no history of prior trauma or surgery. Findings of the examination of right eye were essentially normal. The child had no light perception in the left eye. There was ciliary staphyloma, diffuse corneal edema, dilated fixed pupil, aphakia, and an intraocular pressure of 34 mm Hg by Perkin applanation tonometry under anesthesia. The fundus view was unclear. Ultrasound B-scan showed an increase in the axial length, a clear vitreous cavity, and optic disc cupping Figure 1 A. There was no evidence of subluxation of the crystalline lens or an intraocular mass. An immersion ultrasound B-scan, however, was not done. The glaucoma specialist performed semiconductor diode laser transscleral cyclophotocoagulation to control the intraocular pressure. The child had periodic follow-up at the glaucoma clinic thereafter. The child reported severe pain in the left eye in January and had evisceration by a pediatric ophthalmologist. A repeated ultrasound B-scan of the eye was not performed before evisceration. Histopathology of the eviscerated tissue showed a malignant round cell tumor with areas of necrosis and calcification, based on which a diagnosis of retinoblastoma was made Figure 1 , B and C , and the child was referred to the ocular oncology service. A computed tomographic scan did not reveal optic nerve invasion or orbital extension Figure 1 D. There was no evidence of systemic metastasis. Results of bone marrow biopsy and cerebrospinal fluid cytology were normal. The child was given high-dose chemotherapy with a combination of carboplatin, vincristine, and etoposide for 3 cycles, followed by

eyelid-sparing orbital exenteration Figure 1 E , cGy to convert gray to rad, multiply by fractionated external beam radiotherapy to the orbit, and continued chemotherapy for 12 cycles. The child is alive and well, with no local recurrence or systemic metastasis 41 months after completion of treatment Figure 1 F. View Large Download An 8-year-old child with retinoblastoma who had evisceration of a painful blind eye with the clinical diagnosis of staphyloma with secondary glaucoma after transscleral cyclophotocoagulation case 1. A, The posterior segment is anechoic on ultrasound B-scan performed at the initial visit when the child was aged 3 years. D, A postevisceration axial computed tomographic scan shows an intrascleral implant and no evident orbital retinoblastoma. E, An intraoperative photograph shows eyelid-sparing orbital exenteration. F, Appearance of the exenterated socket is shown at the final follow-up. Case 2 An 8-year-old boy had undergone evisceration elsewhere with a history of painful blind right eye following trauma and a clinical diagnosis of secondary glaucoma. Imaging had not been performed before evisceration. Eviscerated tissue had not been submitted for histopathology by the comprehensive ophthalmologist who had performed the surgery. The child developed a painful, rapidly growing orbital mass 2 months following evisceration Figure 2 A and was referred to our ocular oncology service. A computed tomographic scan showed a large orbital mass with specks of intralesional calcification, suggestive of orbital recurrence of retinoblastoma Figure 2 B. Incisional biopsy of the orbital mass confirmed the diagnosis of retinoblastoma Figure 2 C. The child received high-dose chemotherapy with a combination of carboplatin, etoposide, and vincristine for 3 cycles Figure 2 , D and E , followed by an eyelid-sparing orbital exenteration, cGy fractionated external beam radiotherapy to the orbit, and continued chemotherapy for 12 cycles. The child had no local recurrence or systemic metastasis at 36 months following completion of treatment. View Large Download An 8-year-old child with orbital recurrence of retinoblastoma following evisceration of a painful blind eye with glaucoma secondary to trauma case 2. A, A large fleshy right orbital mass is seen. B, An axial computed tomographic scan demonstrates a large soft-tissue mass in the right orbit with specks of intralesional calcification. D, Following 6 cycles of neoadjuvant chemotherapy, the orbital tumor shows significant resolution. E, A computed tomographic scan axial cut shows significant resolution of the orbital tumor following 6 cycles of neoadjuvant chemotherapy. The right eye had no light perception. The anterior segment showed features of secondary angle-closure glaucoma and complicated cataract. Opaque media precluded fundus view. Ultrasound B-scan showed a large ciliochoroidal mass with low internal reflectivity Figure 3 A. The mass filled the vitreous cavity. Enucleation was advised with the clinical suspicion of a uveal melanoma. The patient was subsequently lost to follow-up. He developed severe pain 1 month later and consulted a comprehensive ophthalmologist elsewhere. The ophthalmologist, who was unaware of prior imaging and clinical diagnosis, proceeded with evisceration of the painful blind eye but did not submit eviscerated tissue for histopathology. Six months later, the patient developed an orbital mass Figure 3 B and was referred to our ocular oncology service. A computed tomographic scan showed a large, isodense orbital mass suggestive of orbital recurrence of uveal melanoma. Systemic evaluation did not reveal any metastasis. The patient had an eyelid-sparing orbital exenteration. Histopathology confirmed the diagnosis of melanoma Figure 3 C. The patient received cGy fractionated orbital external beam radiotherapy. He was alive and well at last visit, 10 months after treatment, with no local recurrence or systemic metastasis. View Large Download A year-old man with orbital recurrence of melanoma following evisceration of a painful blind eye with retinal detachment and secondary glaucoma case 3. A, Ultrasound B-scan with cross vector shows a large intraocular mass filling the entire vitreous cavity top panel , with low internal reflectivity bottom panel , suggestive of uveal melanoma. B, A right orbital mass was noticed 6 months following evisceration. She had earlier undergone cataract surgery, followed by trabeculectomy elsewhere about 10 years ago. She had absent light perception, scarred bleb, and opaque cornea with spheroidal degeneration, precluding further evaluation of the anterior segment and fundus. Ultrasound B-scan showed a closed funnel retinal detachment and no evidence of an intraocular mass. Immersion B-scan, however, was not performed. The patient was advised to have evisceration for symptomatic relief by an oculoplasty surgeon. Histopathology of the eviscerated tissue showed neoplastic cells in glandular pattern with clear cytoplasm, vesicular nucleus, and prominent nucleoli, suggestive of adenocarcinoma of the ciliary body Figure 4. Results of systemic evaluation were unremarkable. The patient

was further treated with enucleation and cGy fractionated orbital external beam radiotherapy. She was alive and well 23 months following completion of treatment. [View Large Download](#) A year-old woman with adenocarcinoma of the ciliary body discovered following evisceration of the painful blind right eye after trabeculectomy case 4. Case 5 An year-old boy had a history of an enlarged left eyeball by birth. He had earlier had a trabeculectomy elsewhere with a diagnosis of developmental glaucoma. He had light perception, intraocular pressure of 34 mm Hg by applanation tonometry, plexiform neurofibroma of the left upper eyelid [Figure 5 A](#) , buphthalmos with scarred bleb, enlarged and edematous cornea, Lisch nodules on the iris, ectropion uveae, and total cataract. Computed tomography of the orbit showed an enlarged left eyeball, and a hypoplastic greater wing of the sphenoid [Figure 5 B](#). There was no evidence of an intraocular mass. The child was diagnosed with neurofibromatosis type 1 with developmental glaucoma, status post trabeculectomy. The child had evisceration of the disfigured left eye and debulking of the eyelid plexiform neurofibroma by an oculoplastic surgeon for cosmetic concern. Histopathology of the eviscerated tissue showed thickened choroid with nests of mature polygonal ganglion cells admixed with nerve fibers [Figure 5 C](#) , focal areas of ganglion cells, and Pacinian bodies [Figure 5 D](#) , diagnostic of choroidal ganglioneuroma. The child was observed. He showed no local tumor recurrence at the final follow-up at 33 months. [View Large Download](#) An year-old boy with neurofibromatosis type 1, with ganglioneuroma of the choroid in the buphthalmic left eye after trabeculectomy, eviscerated for cosmetic concern case 5. [A](#), [A](#) clinical photograph of the face shows plexiform neurofibroma of the left upper eyelid.

## 2: Surgical Management of Intraocular Tumors | Ento Key

*Written by world-renowned authorities in ocular oncology at the Wills Eye Institute, this text/atlas is a comprehensive pictorial and textual guide to the clinical features, pathology, diagnosis, and management of intraocular tumors and pseudotumors.*

Acquired fibrovascular retinal hemangiomatous lesion. Treatment Management decisions should be based on the desire to preserve or restore vision in affected eyes. The most commonly employed method of management is photocoagulation of the vascular tumor, particularly effective against tumor that are less than or equal to 2 mm in attached retina. In successfully treated patients the hemangioma becomes atrophic and involute, the feeding and draining retinal vessels lose their dilation and tortuosity and associated exudates disappear gradually. For peripheral and some larger postequatorial lesions, transconjunctival or trans-scleral cryotherapy can be employed. Treatment must be repeated at intervals of weeks until the lesion totally obliterate and the retinal feeder and drainer vessels are back to normal caliber. Eyes with secondary exudative or tractional retinal detachment associated with large retinal capillary hemangioma or multiple retinal capillary hemangiomas usually require pars plana vitrectomy with membrane peeling and frequently endophotocoagulation of the retinal tumor or tumors. Plaque radiotherapy appears to work well for medium-sized to large retinal capillary hemangiomas located 3 mm or more from the optic disc. Visual prognosis depend on the size, location and number of lesions, the extend of intraretinal and subretinal exudation and the amount of vitreo-retinal fibroplasia that develops in response to the lesion. Most retinal capillary hemangiomas can now be controlled, if not eradicated by local obliterate therapy. Are characterized by the formation of grapelike clusters of thin-walled saccular angiomatous lesions in the inner retina or on the optic nerve head. The blood flow is derived from the retinal circulation and is relatively stagnant, producing a characteristic fluorescein picture. The lesion has no recognized malignant potential. Occasional patients have small hemangiomas and telangiectasia of the skin and similar lesion in the central nerve system. Generally asymptomatic, unless it involves the macula or bleed into the vitreous. Most patients tumor is detected on routine ophthalmic examination. Typical appears as a cluster of vascular saccules within the sensory retina in association with retinal vein of anomalous appearance that is neither dilated or tortuous. The dilated saccular lesions fill slowly during angiography and plasma-erythrocyte layering occurs as a result of the sluggish blood flow; leakage is characteristically absent, correlating with the absence of subretinal fluid and exudate, serving to differentiate it from retinal telangiectasia, von Hippel retina angiomatosis and racemose aneurysm of the retina. Treatment Is usually no indicated unless recurrent vitreous hemorrhage develops, in which case photocoagulation or cryotherapy may be effective. Visual prognosis is good unless the hemangioma involves the macula. Choroidal hemangioma Introduction Is a benign hamartomatous disorder 3A , occurs in two distinct clinical forms: They are relatively uncommon; appear to be vascular birthmarks but their precise incidence in unknown. Diffuse hemangiomas are usually detected at baseline ophthalmic evaluation of patients who have a facial nevus flammeus before the onset of symptoms. The fundus typically has a much more saturated red appearance that on the uninvolved side. The choroid tends to be thickened diffusely by the hemangiomatous vascular lesion, but accentuated and sometimes nodular thickening occurs frequently in the macular and circunpapillary regions. The choroidal thickening around the optic disc commonly results in prominent disc cupping that resembles glaucomatous optic neuropathy. Elevated intraocular pressure usually is caused by elevated episcleral and orbital venous pressure, angle malformation or both; is a feature of many eyes that have diffuse choroidal hemangioma. The retinal pigment epithelium that overlies ticker portions of a diffuse choroidal hemangioma often undergoes fibrous metaplasia, gives a whitish appearance to the part of the lesion. Serous retinal detachment is an eventual complication in many eyes that have diffuse choroidal hemangioma. Typical lesion ranges from 3 to 7 mm in diameter and 1 to 3 mm in thickness. Almost all are located within two disc diameters from the optic disc, foveola or both at their posterior margins. The retinal pigment epithelium that overlies the lesion commonly undergoes degenerative changes, including fibrous metaplasia and occasionally degenerative calcification. Serous retinal detachment occurs frequently as a

complication, and can be mistaken for central serous retinopathy. Fluorescein angiography of circumscribed choroidal hemangiomas typically reveals very early hyperfluorescence of larger-caliber choroidal blood vessels either before or simultaneously with the initial filling of the retinal arterioles. By late frames, fluorescein commonly stains the entire lesion and any associated subretinal fluid. Optical coherence tomography can highlight and document the subretinal fluid accumulation along with cystic retinal degeneration that can occur overlying the lesion. The full extent of a circumscribed choroidal hemangioma is usually revealed much more clearly. In a diffuse choroidal hemangioma characteristically reveals moderately bright generalized posterior choroidal thickening and prominent optic disc cupping. Ultrasonography also is usually to detect serous retinal detachment. Benign reactive lymphoid hyperplasia of choroid, leukemic choroidal infiltration, diffuse posterior scleritis and uveal effusion syndrome. Treatment The treatment is directed toward limitation or reversal of visual loss related to secondary retinal detachment, glaucoma or other complications. Some asymptomatic cases can be observed. Photodynamic therapy PDT using verteporfin has been used with considerable success as treatment of small to medium-size circumscribed choroidal hemangiomas and is currently regarded as the treatment of choice for such lesions in many centers. In some cases PDT has been combined with intravitreal anti-vascular endothelial growth factor drug therapy to treat eyes with a large amount of turbid subretinal fluid, subfoveal tumor location, or both. In patients who have an extremely thick choroidal hemangioma, extensive non-rhegmatogenous retinal detachment, or a diffuse or circumscribed choroidal hemangioma that failed to respond to PDT. Low dose ocular irradiation appears to be an effective therapeutic option. Several different radiation therapy methods external beam photon radiotherapy, plaque radiotherapy, proton beam irradiation, gamma knife radiotherapy and stereotactic radiotherapy have been employed with good success in selected patients. Radiation therapy induce partial or total tumor regression, stimulate gradual reabsorption of subretinal fluid that is usually sustained for many months to years, and preserve useful vision in at least some affected eyes. Is the macula involved progressive degeneration of the overlying retinal pigment epithelium and sensory retina often occurs, leading to progressive visual loss. In addition, secondary serous retinal detachment can cause profound visual impairment. Chronic bullous retinal detachment is frequently followed by neovascularization of the iris and neovascular glaucoma. Following PDT prompt clinical regression of the tumor generally occurs, associated subretinal fluid usually disappears promptly within 3 months. Sustained local tumor regression and lack of reaccumulation of subretinal fluid have been reported in most but not all cases. Following radiation therapy, the hemangioma generally undergoes partial regression and the subretinal fluid slowly goes away, but may take up to 6 months or more. In the great majority of eyes treated by radiation therapy the subretinal fluid never reaccumulates. The low dose of radiation used to treat such tumors rarely causes side effects. Retinal arterio-venous malformation Wyburn-Mason Syndrome Introduction Is a rare sporadic disorder characterized by congenital arterio-venous malformation in which no intervening capillary bed exists racemose angioma principally of the retina and brain. Lesions are typically unilateral, nonhereditary and located in the retina or optic nerve. The diagnosis is most commonly made later in childhood, often detected as an incidental finding in an asymptomatic patient or as cause of visual impairment in an amblyopic eye. Abnormal capillary plexus between the major vessels of the arterio-venous malformations. Arteriovenous malformations lack any intervening capillary between the artery and vein. Arteriovenous malformations are the most extensive with dilated and tortuous vessels and no apparent distinction between the artery and vein. If the lesions are larger they may be associated with subretinal fluid and exudate. It demonstrates abnormal arteriovenous connections and presence<sup>34</sup> or absence of intervening capillaries. In the most severe cases Grade III arteries and veins cannot be differentiated even on angiography. Abnormal retinal vasculature characteristically demonstrates absence of leakage. Macular fibrosis may lead to visual loss. Small peripheral vasoproliferative tumors, lacking significant exudate or maculopathy may be managed by periodic observation. Other treatment options include plaque brachytherapy, laser photocoagulation and photodynamic therapy. Gass JD, Braunste in R. Sessile and exophytic capillary angiomas of the juxtapapillary retina and optic nerve head. Clinical features and natural history of von Hippel Lindau disease. Cavernous hemangioma of the retina. Vitreoretinal surgery for severe retinal capillary hemangiomas in von Hippel Lindau disease. Hardwig P, Robertson OM. Presumed

acquired retinal hemangiomas. Benefits and complications of photodynamic therapy of papillary capillary hemangiomas. Long-term results of laser treatment for retina angiomatosis in von Hippel. Saudi Journal of Ophthalmology, Volume 21, No. Eur J Med Res ;5: Direct and feeder vessel photocoagulation of retinal angiomas with dye yellow laser. The recognition and treatment of early angiomatosis retinae and use of cryosurgery as an adjunct to therapy. Trans Am Ophthalmol Soc ; Retinal capillary hemangioma treated with verteporfin photodynamic therapy. Am J Ophthalmol ; A neurooculo- cutaneous syndrome. Nine cases of cavernous hemangioma of the retina. American Journal of Ophthalmology ; Familial cavernous malformations of the central nervous system and retina. Acta Neurol Scand ; Vitrectomy for persistent vitreous hemorrhage from cavernous hemangioma of the optic disc. A four-generation pedigree with neurocutaneous manifestations and an example of bilateral retinal involvement. Witschel H, Font RL. Hemangioma of the choroid. A clinicopathologic study of 71 cases and a review of the literature. Survey of Ophthalmology ; The ocular manifestations of the Sturge-Weber syndrome. Photodynamic therapy of circumscribed choroidal haemangioma. Br J Ophthalmol ;

**3: Intraocular Tumors: An Atlas and Textbook - Jerry A. Shields, Carol L. Shields - Google Livros**

*This is a retrospective, nonrandomized, clinicopathological case series of patients with previously unsuspected intraocular tumors diagnosed by histopathology of eviscerated blind eyes.*

Histopathological report Findings from histopathological evaluation revealed an intact conjunctival epithelium. Focally, the epithelium had areas of primary acquired melanosis with cellular atypia. In the subepithelial tissue, nests of slightly pigmented, uniform-looking tumor cells were surrounded by a chronic, nonspecific lymphoplasma-cellular infiltration. Nests of tumor cells were seen within the sclerocorneal graft as well as inside the underlying cornea, reaching almost to the middle of the cornea. The chamber angle showed neovascularization and was partly invaded by tumor cells growing on the corneal and iridal surfaces with invasion of iris and trabecular meshwork. The ciliary body was partially thickened with collateral retinal detachment. There was a diffuse invasion of the ciliary body by mainly epithelioid tumor cells Figure 4. Single sections showed tumor cells in the scleral emissaria connecting the tumor cell masses inside the sclera and the cornea with the ciliary body Figure 5. The choroid, attached retina, and optic nerve appeared normal. All different parts of the tumor of conjunctiva, cornea, sclera, and ciliary body expressed the same immunohistochemical-staining pattern positive for S antigen and HMB antigen and showed polygonal nuclei, prominent nucleoli, and a small cytoplasmic rim. Comment Intraocular extension of a conjunctival malignant melanoma is an extremely rare entity. Therefore, the coincidental finding of epibulbar and intraocular pigmented lesions in our patient primarily suggests a uveal melanoma with extraocular extension. The clinical and histopathological appearance of the ciliary body changes especially seemed to indicate a primary epithelioid malignant melanoma of the ciliary body Figure 4. The presumed ciliary body tumor showed collateral retinal detachment and invasion of chamber angle, trabecular meshwork, iris, cornea, and sclera and, most likely, extraocular extension. Indeed, there is no way to directly prove whether the tumor in our patient was originating from the conjunctiva or the ciliary body because there is no method to differentiate between conjunctival or uveal melanocytes. However, we present 6 arguments supporting our hypothesis of a primary conjunctival melanoma with intraocular extension. This finding is not consistent with an epibulbar growth originating from a primary intraocular tumor. This seems to represent the pathway of intraocular invasion. Additionally, there are no exact data concerning the radiosensitivity of the conjunctival malignant melanoma and the advantages of an adjuvant radiotherapy. Nevertheless, there was recurrent tumor growth with partially deep scleral and corneal extension. This extension of the tumor led to a deep excision by lamellar sclerokeratectomy. Despite treatment, tumor cells persisted deep inside the sclera and cornea underneath the transplant and were able to invade the eye. Compared with the technique published by Shields et al,<sup>8</sup> the deep scleral extension of the tumor in our patient demanded deeper scleral incisions of 0. Perhaps the preceding limbal surgery with partial removal of the Bowman membrane in our patient reduced the natural barrier against intraocular tumor invasion, as discussed by Gow and Spencer. This could indicate that the scleral and corneal barrier is normally resistant to penetration by an epibulbar malignant melanoma but may lose this function after lamellar surgery. Accepted for publication November 10,

## 4: Options for management of intra ocular tumors

*The management of intra ocular tumors has undergone a sea change from the era of enucleation or external beam radiation. With the advent of new chemotherapy protocols, globe and vision salvage have become possible in a majority of cases of retinoblastoma. This article is an overview of the various.*

Management of Ocular Surface Tumors: Karp Abstract OSSN encompasses a range of corneal and conjunctival lesions from intraepithelial dysplasia to invasive squamous cell carcinoma. The mainstay of treatment for OSSN has traditionally been surgical excision with wide margins and cryotherapy. Increasing evidence on the efficacy and safety of medical therapy and the avoidance of surgical complications has made topical chemotherapy increasingly popular among corneal specialists. The most common topical agents used for the treatment of OSSN include mitomycin C, 5-fluorouracil, and interferon  $\alpha$  2b. Herein, we review recent advances in the surgical and medical management of OSSN and discuss advantages and disadvantages of each approach. The role of ultra high-resolution optical coherence tomography in the diagnosis and treatment of primary and recurrent OSSN lesions is also discussed. Keywords ocular surface squamous neoplasia; interferon  $\alpha$ 2b; mitomycin C; 5-fluorouracil; ultra-high-resolution optical coherence tomography. Clinical survey of melanocytic and nonmelanocytic conjunctival tumors. Conjunctival lesions in adults. A clinical and histopathologic review. Ocular surface squamous neoplasia. A review of the etiology of squamous cell carcinoma of the conjunctiva. Incidence of ocular surface epithelial dysplasia in metropolitan Brisbane. Epidemiology of squamous cell conjunctival cancer. Cancer Epidemiol Biomarkers Prev. Effect of ambient solar ultraviolet radiation on incidence of squamous-cell carcinoma of the eye. Risk factors in the development of ocular surface epithelial dysplasia. Basti S, Macsai MS. Ocular surface squamous neoplasia: Association of human papilloma virus with pterygia and ocular surface squamous neoplasia. A possible marker for human immunodeficiency virus infection? Ocular involvement in xeroderma pigmentosum. Surgical management of conjunctival tumors. The Lynn B. Conjunctival and corneal intraepithelial and invasive neoplasia. Late recurrences and the necessity for long-term follow-up in corneal and conjunctival intraepithelial neoplasia. Topical mitomycin C for the treatment of conjunctival and corneal epithelial dysplasia and neoplasia. Mitomycin C treatment for conjunctival-corneal intraepithelial neoplasia: Preoperative topical and intraoperative local mitomycin C adjuvant therapy in the management of ocular surface neoplasias. Topical mitomycin-C for partially excised conjunctival squamous cell carcinoma. The intraoperative use of mitomycin-C in excision of ocular surface neoplasia with or without limbal autograft transplantation. Topical mitomycin C for extensive, recurrent conjunctival-corneal squamous cell carcinoma. Chemoreduction with topical mitomycin C prior to resection of extensive squamous cell carcinoma of the conjunctiva. Randomized controlled trial of topical mitomycin C for ocular surface squamous neoplasia: Topical application of 5-fluorouracil in premalignant lesions of cornea, conjunctiva and eyelid. Topical 5-fluorouracil in treating epithelial neoplasia of the conjunctiva and cornea. Conjunctival squamous cell carcinoma treated with topical 5-fluorouracil. Regression of limbal epithelial dysplasia with topical interferon. Interferon treatment for corneolimbal squamous dysplasia. Perilesional and topical interferon  $\alpha$ -2b for conjunctival and corneal neoplasia. Paul S, Stone DU. Intralesional bevacizumab use for invasive ocular surface squamous neoplasia. J Ocul Pharmacol Ther. Refractory squamous cell carcinoma of the conjunctiva treated with subconjunctival ranibizumab Lucentis: Ophthal Plast Reconstr Surg. Topical retinoic acid in dysplastic and metaplastic keratinization of corneconjunctival epithelium. Treatment of conjunctival and corneal epithelial neoplasia with retinoic acid and topical interferon  $\alpha$ -2b: Gupta A, Muecke J. Treatment of ocular surface squamous neoplasia with Mitomycin C. Khong JJ, Muecke J. Complications of mitomycin C therapy in eyes with ocular surface neoplasia. Long-term results of topical mitomycin C 0. Limbal stem cell deficiency following topical mitomycin C treatment of conjunctival-corneal intraepithelial neoplasia. Stability of mitomycin admixtures. Am J Hosp Pharm. Punctal-canalicular stenosis related to systemic fluorouracil therapy. Redefining the role of interferon in the treatment of malignant diseases. Pegylated interferon  $\alpha$  2b for treatment of ocular surface squamous neoplasia: Topical interferon  $\alpha$  2b eye-drops for treatment of ocular surface squamous

neoplasia: Treatment of conjunctival and corneal intraepithelial neoplasia with topical interferon alpha-2b. Regression of presumed primary conjunctival and corneal intraepithelial neoplasia with topical interferon alpha-2b. Treatment of recurrent corneal and conjunctival intraepithelial neoplasia with topical interferon alfa-2b. Long-term follow-up of conjunctival and corneal intraepithelial neoplasia treated with topical interferon alfa-2b. Topical interferon or surgical excision for the management of primary ocular surface squamous neoplasia. Successful management of conjunctival intraepithelial neoplasia by interferon alpha-2b. Huerva V, Mangues I. Treatment of conjunctival squamous neoplasias with interferon alpha 2ab. Topical interferon alfa-2b for management of ocular surface squamous neoplasia in 23 cases: Predictors of ocular surface squamous neoplasia recurrence after excisional surgery. Diagnosis and management of conjunctival and corneal intraepithelial neoplasia using ultra high-resolution optical coherence tomography. Ultra-high resolution optical coherence tomography for differentiation of ocular surface squamous neoplasia and pterygia. Ultra highresolution anterior segment optical coherence tomography in the diagnosis and management of ocular surface squamous neoplasia. A Comparison of Recurrences and Complications.

## 5: Eye tumors: an overview

*Ocular surface tumors are rare but potentially deadly diseases of the conjunctiva and/or cornea. It is important for ophthalmologists to recognize the characteristics of ocular surface tumors and to have an understanding of their management.*

It consists of a broad range of pathologic squamous cell dysplasia including: There is greater incidence near the equator. Most lesions are pathologically benign, such as papillomas or actinic keratosis. However, other lesions are more nefarious, such as carcinoma in situ and invasive squamous cell carcinoma. OSSN appears on slit lamp biomicroscopy as a poorly defined gelatinous lesion, usually blending with surrounding conjunctiva. There is typically an abrupt transition from normal to dysplastic epithelium. Feeder vessels often supply invasive masses, and as the lesion becomes more advanced, there is decreasing mobility of the tumor due to the conjunctiva becoming fixed to the deeper sclera. It should be noted that carcinoma in situ and invasive squamous cell carcinoma can be very difficult to differentiate at the slit lamp. Thus, biopsy is very helpful in making the diagnosis. Conjunctival squamous cell carcinoma in situ Slit lamp photograph displaying patchy, limbal-based gelatinous mass. Vascular engorgement is noted. This is an example of carcinoma in situ. Pathology The gold standard for diagnosis is histopathological evaluation following biopsy. Carcinoma in situ is characterized by full-thickness replacement of the epithelium with anaplastic cells; however, the basement membrane remains intact and the underlying substantia propria is not affected. Histology displays a mixture of spindle and epidermoid cells, with disorganization of cells, increased nuclear to cytoplasm ratio, and abnormal polarity. There is generally a characteristic demarcation between diseased epithelium and adjacent normal tissue Figure As the tumor invades, it can appear as cords of invasive cells or as broad, expansive fronds. Image on right displays acanthosis, dyskeratosis, and bizarre cells black arrow. Treatment Adequate treatment of squamous cell neoplasms depends upon the clinical characteristics of the lesions. Factors such as size, location, and invasiveness of the tumor influence the appropriate treatment of neoplasms. For discrete masses, complete excision with adequate margins is the treatment of choice, often aided by alcohol epitheliectomy. It is important to ensure adequate margins, which may include up to mm of uninvolved conjunctiva. Most surgeons will apply double freeze thaw cryotherapy to the adjacent bulbar conjunctiva in order to provide better local control. Additionally, amniotic membrane grafts can be used to close a resection site. No consensus exists for use of these therapies. Enucleation or exenteration may be required with invasive squamous cell carcinoma. Enucleation is necessary in cases of invasion through the cornea or sclera without orbital or regional spread. Exenteration is necessary when tumor has invaded the orbit. Benign tumors are reactive lymphoid hyperplasia, intermediate lesions present as atypical lymphoid hyperplasia, and malignant tumors are lymphomas. Lymphoid tumors of the conjunctiva are very rare, and the exact incidence is unknown. In a large series of conjunctival tumors, Shields et al. Known risk factors for malignancy include older age, history of systemic lymphoma, and immunosuppression. There is a predisposition for the inferior fornix. Thus, excisional biopsy is required for diagnosis. Conjunctival benign lymphoid hyperplasia Slit lamp photo displaying superior conjunctival lesion with characteristic salmon patch appearance. Slit lamp photo displaying salmon patch lesion extending into the semilunaris and caruncle. Pathology It is important to obtain immunofluorescence testing and fresh samples for appropriate pathological diagnosis. Benign reactive lymphoid hyperplasia is characterized by polymorphic, well-differentiated lymphocytes and possibly plasma cells. These cells tend to have well-developed germinal centers. Lymphomas are monomorphic, without germinal centers. Flow cytometry is helpful in determining whether cells are monoclonal or polyclonal, whether cells are B or T cells, and if DNA abnormalities are present. In a patient with a suspected lymphoid tumor, a complete medical workup for systemic lymphoma is warranted. Included in the initial investigation is a history and physical, complete blood count with differential, and magnetic resonance imaging MRI of brain and orbits, as well as PET scan to identify a systemic lymphoma. In addition to a comprehensive evaluation, excisional biopsy should be completed to identify the underlying pathology. In the case of benign reactive lymphoid hyperplasia, treatment can be observation or topical steroids for a few weeks. In the case of

low-grade lymphoid neoplasm, low-dose external beam radiation therapy is recommended. In the case of high-grade lymphoma, higher dose external beam radiation is recommended usually Gy. If systemic lymphoma is discovered on workup, then treatment consists of chemotherapy with or without radiation. Most localized lymphomas respond well to excision and radiation. In patients with systemic lymphoma, chemotherapy is often indicated for treatment, with rituximab as the treatment of choice. *Surv Ophthalmol* ;42 4: Uveal and conjunctival malignant melanoma in Denmark, *Ophthalmic Epidemiol* ;12 4: Incidence of noncutaneous melanomas in the U. *Arch Ophthalmol* ; Meta-analysis of risk factors for cutaneous melanoma: Family history, actinic damage and phenotypic factors. *Eur J Cancer* ;41 Management of conjunctival malignant melanoma: *Expert Rev Ophthalmol* ;9 3: *Arch Dermatol* ; 8: *Am J Ophthalmol* ; 6: Primary acquired melanosis of the conjunctiva: *Trans Am Ophthalmol Soc* ; Clinical characterization of primary acquired melanosis. *Invest Ophthalmol Vis Sci* ;36 8: Conjunctival naevi in Denmark A year follow-up study. *Acta Ophthalmol Scand* ;74 4: Benign conjunctival melanocytic lesions. Clinicopathologic characteristics of premalignant and malignant melanocytic lesions of the conjunctiva. Surgical management of circumscribed conjunctival melanomas. *Ophthal Plast Reconstr Surg* ;14 3: *Can J Ophthalmol* ;39 4: Regional lymph node assessment for conjunctival melanoma: *Br J Ophthalmol* ;92 4: Ocular surface squamous neoplasia. *Surv Ophthalmol* ;39 6: The aetiology and associations of conjunctival intraepithelial neoplasia. *Br J Ophthalmol* ;90 1: Demographics, etiology, and behavior of conjunctival squamous cell carcinoma in the 21st century. A histopathologically based retrospective study. *Acta Ophthalmol Scand* ;78 6: Tumors of the conjunctiva and cornea. *Surv Ophthalmol* ;49 1: Intraepithelial and invasive squamous cell carcinoma of the conjunctiva: *Br J Ophthalmol* ;83 1: Ocular surface squamous neoplasia - Review of etio-pathogenesis and an update on clinico-pathological diagnosis. *Saudi J Ophthalmol* ;27 3: *Expert Rev Ophthalmol* ;12 1: *Asia Pac J Ophthalmol Phila* ;6 2: Tumors of the ocular surface: *Indian J Ophthalmol* ;63 3: Late recurrences and the necessity for long-term follow-up in corneal and conjunctival intraepithelial neoplasia. Clinical survey of melanocytic and nonmelanocytic conjunctival tumors. Treatment of conjunctival lymphomas. *Semin Ophthalmol* ;20 4: Lymphoproliferative lesions of the ocular adnexa. Analysis of cases.

## 6: Intraocular Vascular Tumors - EyeWiki

*Epidemiology and risk factors. Conjunctival melanoma is a rare ocular tumor comprising about 5% of ocular melanomas and % of all melanomas.[1,90] Conjunctival melanoma arises from melanocytes among the basal cells of the conjunctival epithelium.*

New international classification for retinoblastoma Pathology Retinoblastomas are characterized histopathologically by basophilic cells with minimal cytoplasm surrounding a lumen in a rosette formation or radially arranged around a central tangle of fibrils in a pseudorosette formation. Often times, necrosis and hemorrhage are present within the tumors, as they tend to outgrow their vascular supplies. Endophytic retinoblastoma lesion in the right eye obscuring the view of the macula and optic disc. Cytogenetics Linkage analysis and deletion techniques discovered the RB1 to be localized to chromosome 13q14 Yunis and Ramsay, The gene spans kb, is composed of 27 exons, and encodes a 4. The resulting protein product is a kD nuclear phosphoprotein, consisting of amino acids. Genes The RB1 gene encodes a protein that is a regulator at the major checkpoint of the cell cycle, between the G1 and S-phase. In its normal non-phosphorylated form, the retinoblastoma protein pRB binds to transcriptional factors, like E2F, to prevent entry into the S-phase. The phosphorylated form of pRB dissociates from E2F, which allows the transcription factor to bind DNA and promote progression through the cell cycle. Abnormal RB1 function allows for continuous entry into the S-phase and rapid cell division, which causes tumor formation. Treatment Treatment of retinoblastoma is often performed in a multi-modal approach including enucleation, external beam radiation, chemotherapy, transpupillary thermotherapy, cryotherapy, and brachytherapy. The dosing regimens, schedules, and chemotherapeutic agents vary significantly among oncology centers, but the most common includes carboplatin, etoposide and vincristine. Most centers use chemotherapy to reduce the tumors, which allows focal treatment like laser and cryotherapy to be more effective. Enucleation is still used in very advanced cases. Some centers use periocular carboplatin with systemic chemotherapy to salvage globes with advanced disease. Anti-angiogenic agents and intra-arterial chemotherapy infusions are also currently being investigated. Prognosis The most significant factor leading to death is extraocular invasion by the tumor, with a considerable delay in initial diagnosis also contributing to a reduced likelihood of survival. Disease Medulloepithelioma Etiology Medulloepithelioma is a tumor that arises from the epithelium of the medullary tube, most often the ciliary body. It can take a teratoid or nonteratoid form and usually presents itself as a unilateral congenital disease, although bilateral, juvenile, and adult-onset cases have also been reported. Broughton and Zimmerman, Epidemiology Medulloepithelioma is a rare tumor with an incidence of 1 case per - people Augsburger and Schneider, Medulloepithelioma has no racial and gender predilection, has no clear pattern of inheritance, and no identifiable risk factors. The mean age at diagnosis is 4 - 5 years. Clinics It most commonly presents as a gray-white lesion of the anterior chamber angle, but can present as a diffuse mass causing leukocoria Broughton and Zimmerman, Neovascular glaucoma in a child with a normal posterior segment, iris notching, and an unexplained cyclitic membrane are features that may assist in diagnosis. While clinical examination may be sufficient to diagnose medulloepithelioma, ultrasound can be a useful adjunct as it demonstrates cystic spaces and a lack of calcifications. Pathology Histopathologic analysis reveals a tumor composed of epithelium that can be arranged in cords and sheets separated by cystic spaces containing proteinaceous material. Teratoid forms contain heterotopic elements including skeletal muscle and cartilage. Malignant tumors often have areas consisting of poorly differentiated neuroblastic cells, increased mitotic activity, sarcomatous areas, and tumor invasion of other ocular tissue, regardless of extraocular extension. Medulloepithelioma with opaque white lesion. Treatment Currently, there is not a definitive treatment for medulloepithelioma. Observation is often recommended for smaller tumors without sequelae. Primary enucleation is recommended if the tumor is large, there is extrascleral extension, the eye is blind and painful, or there is neovascular glaucoma. Local resection and invasive diagnostic procedures should be avoided as they may lead to recurrence, direct seeding of the orbit, or creation of a tract for subsequent tumor migration from the globe and into the orbit. Prognosis The natural history of untreated medulloepitheliomas is

essentially unknown. Metastasis in medulloepithelioma is very rare, but portends a more negative prognosis when present. Only lesions with malignant features metastasize, but neither teratoid features nor malignant morphology predict mortality. While there has been no reported deaths or metastases in patients who undergo definitive enucleation without prior diagnostic invasive procedures, patients who have had prior invasive procedures are thought to have a higher mortality rate. FAP is a syndrome with an autosomal dominant mode of inheritance that causes the development of hundreds of pre-malignant colonic polyps and is caused by a mutation in the APC gene located on chromosome 5q. Not only are POFLs bilateral, numerous, and pisciform in shape, but they also differ from CHRPE lesions histopathologically as they are shown to have hamartomatous changes in addition to RPE hypertrophy and hyperpigmentation. Etiology Congenital hypertrophy of the retinal pigment epithelium CHRPE is an isolated sporadic congenital lesion with no known underlying genetic basis. Epidemiology While the prevalence of CHRPE is unknown because it usually presents asymptotically, one study demonstrated a prevalence of 1. Age is not a relevant factor in the development of CHRPE because it is a congenital lesion, but studies have shown a median age of diagnosis to be 45 Shields et al. Clinics Because CHRPE are most commonly found in the peripheral retina, patients are commonly asymptomatic and present with round, darkly pigmented, flat lesions that can be surrounded by a hypopigmented halo Lloyd et al. Fundoscopic examination showing classical features of the lesion are sufficient for diagnosis, and no further ancillary studies are needed. The overlying photoreceptor layer may be atrophic, while the underlying choroid and choriocapillaris are usually normal. Congenital hypertrophy of the retinal pigment epithelium located in the superotemporal quadrant of the right eye. Note the flat, round, and darkly pigmented classical appearance of the lesion. Treatment Treatment is usually unnecessary except for the rare instance in which neovascularization presents at the periphery of the lesion. On a rare occasion, CHRPE may transform to malignant adenocarcinoma, but the etiology and most appropriate management has yet to be determined Shields et al. Disease Combined hamartoma of the retina and retinal pigment epithelium RPE Etiology Combined hamartoma of the retina and retinal pigment epithelium RPE is a rare developmental disorder caused by benign proliferations of both the retina and RPE. There may be a systemic association with neurofibromatosis type I and neurofibromatosis type II Palmer et al. They are considered to be congenital lesions in most instances but acquired lesions have been reported infrequently Ticho et al. Epidemiology The prevalence of this occurrence has not been established, but whites have been shown to be more frequently affected Shields et al. Two major studies have shown a mean age of diagnosis years Font et al. Clinics Classically, combined hamartomas present as unilateral, dark, solitary lesions that are slightly elevated with varying amounts of retinal and epiretinal tissue centrally causing progressive traction and vascular tortuosity Font et al. They are located in the macular and extramacular region at equal frequencies Shields et al. Indirect ophthalmoscopy is often sufficient for diagnosis, but fluorescein angiography FA is a useful adjunct as it shows blocking of choroidal perfusion and progressive hyperfluorescence in the late phase Schachat et al. Pathology Histopathologically, combined hamartomas demonstrate infiltration of hyperplastic RPE into the retina and inner retinal surface. Gliosis is significant and is responsible for the tractional changes and vascular tortuosity. Combined hamartoma of the retina and retinal pigment epithelium positioned on the optic disk and adjacent retina with a predominantly glial component. A, Color fundus photography with peripheral areas of traction corresponding to the posterior hyaloid face. B, Fluorescein angiography of the lesion in the same patient demonstrating the striking vascular abnormalities. Treatment While most cases of combined hamartoma are isolated from systemic findings, patients who have been diagnosed should undergo evaluation to exclude neurofibromatosis. Amblyopia therapy has been shown to improve vision in some patients with combined hamartoma Schachat et al. Vitrectomy and membrane removal has been performed, but the visual acuity does not always improve significantly and membranes may recur Shields et al. Consequently, the role of vitrectomy in managing combined hamartoma has not been fully established. Rarely, choroidal neovascularization may occur as a complication and can be treated with laser. Prognosis Combined hamartoma can cause significant visual loss, with a visual acuity Disease Congenital melanocytosis Etiology Ocular melanocytosis is a congenital hyperpigmentation of the globe caused by increased numbers of melanocytes. There is often dermal involvement due to the failure of melanocytes of neural crest cell origin to

reach the intended surface positions, which gives rise to hamartomatous nests in the distribution of the first and second branches of the trigeminal nerve Zaihosseini et al. Epidemiology Ocular melanocytosis is an uncommon condition with a prevalence rate of 0. There are no differences in frequency based on laterality or gender. Clinics Characterized by melanotic pigmentation of the iris, patches of gray-brown scleral pigmentation, and ipsilateral choroidal hyperpigmentation Rahman et al. There may or may not be involvement of the periorbital facial tissues in the distribution of the trigeminal nerve. Diagnosis is based on presentation with classic findings noted during slit-lamp examination. Pathology Histopathologically, ocular melanocytosis is characterized by the presence of dendritic melanocytes in the areas of hyperpigmentation. Congenital melanocytosis revealing gray-brown hyperpigmentation of the sclera and periorbital area of the right eye. Treatment In and of itself, congenital ocular melanocytosis is a benign condition that does not require treatment. However, in the white population there has been shown to be a lifetime risk of developing uveal melanoma of 1 in , which is significantly greater than the 1 in risk observed without underlying congenital melanocytosis Shields et al. Because of the association with uveal melanoma, annual ophthalmic follow-up is recommended for all patients with ocular melanocytosis Rahman et al. Prognosis Visual impairments that arise in the context of ocular melanocytosis are due to the development of complications including uveitis, glaucoma, and cataract. Additionally, the association with an increased frequency of uveal melanoma in the affected eye, has further implications on morbidity and mortality. Disease Uveal nevus Etiology Uveal nevi are stromal, hamartomatous clusters consisting of atypical melanocytes of neural crest origin much like cutaneous melanocytes. They have been described predominantly in three locations including the iris, ciliary body, and choroid. Choroidal nevi have a reported prevalence rate that ranges from 0. They often become pigmented or develop within the first three decades of life, and there is no conclusive data to show an association with gender. Clinics Iris nevi are typically solitary, circumscribed lesions located in the lower quadrants of the iris, ranging from tan to dark brown. Ciliary body nevi present as dome-shaped masses without intrinsic vascularity. Diagnosis of both iris and ciliary body lesions can often be made based on anterior segment evaluation with gonioscopy. Ultrasound biomicroscopy assists in diagnosis by determining size, extent and solid or cystic consistency Conway et al. Choroidal nevi usually do not cause symptoms and present as grayish brown lesions with minimal thickness, and diagnosis can be made by ophthalmoscopy. Pathology Nevi are known to consist of four different cell types including plump polyhedral, slender spindle, intermediate, and balloon cells. Posteriorly, choroidal nevi have been shown to involve full thickness of the choroid with sparing of the choriocapillaris. Pigmented choroidal nevus situated temporal to the macula with overlying drusen. Treatment Periodic ophthalmic examination for nevi to check for growth or malignant progression is recommended for nevi in all locations. For choroidal nevi, follow-up is especially important if the nevi cause decreased vision or visual field defects or have high-risk characteristics such as thickness greater than 2mm, posterior location, orange pigment, subretinal fluid, and absence of drusen Singh et al. Disease Uveal melanoma Etiology Uveal melanoma is the most common primary intraocular malignant tumor, and they originate from the iris, ciliary body, or choroid. The majority of these tumors arise in the choroid, and the predisposing factors include family history of choroidal melanoma, dysplastic nevus syndrome , xeroderma pigmentosum , and congenital ocular melanocytosis. While some arise de novo Sahel et al.

## 7: Clinical Rotations | UC Davis Eye Center

*Surgical management, in the cases of suspicious nevi and focal melanoma, involves excision of the entire melanocytic lesion with wide surgical margins, approximately 3 to 4 mm of normal conjunctiva where possible.*

The selected management depends on factors like tumor size, location, and activity as well as the status of the opposite eye and the age, general health, and psychological status of the patient. Each patient should have a detailed ophthalmic evaluation and the size and extent of the tumor carefully documented with accurate drawings and photography. The known risk factors for growth and metastasis should be considered and the patient should be counseled as to the therapeutic options. Historically, enucleation of the affected eye was once considered to be the only appropriate management for the patient with a posterior uveal melanoma. Several years ago, however, some authorities challenged the effectiveness of enucleation for preventing metastatic disease and even proposed that enucleation may somehow promote or accelerate metastasis. The two most frequently employed treatment methods today are enucleation and plaque radiotherapy. Further specific data on the COMS is listed below under surgical management. More recently, choroidal melanoma has been detected at earlier stages using published risk factors designed to allow detection as early as possible. It has been shown that melanoma prognosis depends on many factors, and tumor thickness is one of the most important. Medical therapy Currently there is no effective oral or intravenous medical therapy for ciliary body or choroidal melanoma. There are tumors that are amenable to therapy in the office setting without operating room intervention. These include periodic observation, laser photocoagulation, transpupillary thermotherapy, and photodynamic therapy. Choroidal nevi often present with overlying retinal pigment epithelial atrophy and drusen signifying a chronic condition. It is estimated that 1 in 10 choroidal nevi evolve into choroidal melanoma. There remains a gray zone of borderline, possibly at-risk tumors measuring 3 mm or less in thickness. Controversy regarding whether these small lesions represent small melanoma versus nevi is a concern. Individual and combined impact of these risk factors is listed in Table 2. A later analysis of choroidal nevus transformation into melanoma in cases revealed similar risk factors but 3 new factors were identified including ultrasound hollowness, absence of drusen, and absence of surrounding halo. Table 3 Documented growth of a melanocytic choroidal tumor is suggestive that the lesion is a choroidal melanoma. Table 4 [14] Based on the few patients with medium size choroidal melanoma who refuse treatment and are followed, natural history studies have found that there is greater mortality and higher risk of death. Studies have shown that xenon achieved better tumor control but argon was associated with fewer complications. TTT is typically delivered in 3 sessions and, at completion, leaves an atrophic chorioretinal scar at the site of the previous tumor. Tumors at the optic disc show greater recurrence and are best managed with plaque radiotherapy combined with thermotherapy. Choroidal melanoma treated with TTT should be followed long-term as delayed recurrence, even with extrascleral extension, can occur. Currently, TTT is used most frequently as a supplement to plaque radiotherapy. In Shields and associates analyzed the clinical factors predictive of tumor recurrence and poor visual outcome following 3 consecutive sessions of primary TTT for choroidal melanoma in patients. Factors predictive of recurrence were optic disc overhung by tumor and increasing number of TTT sessions implying poor response. In Pan et al. In Aaberg et al. These tumors were mostly treated with one session initially, and further treatments were applied as necessary. A particularly bothersome finding was the presence of extraocular extension in 11 cases. Most clinicians prefer TTT only for small and not medium-sized melanoma and most patients receive 3 sessions, even if the tumor appears regressed. The most common side effects of TTT for small melanoma include branch retinal vein occlusion, retinal traction, and retinal hemorrhage. When considering this treatment, it is important to evaluate whether the patient will be able to sustain careful, long-term follow-up to monitor for recurrences. The ideal tumor characteristics for primary TTT include a tumor with dark pigmentation for best diode absorption, a tumor of small size thickness less than 3. The greatest advantage of TTT over plaque radiotherapy is the preservation of vision, and this treatment is particularly beneficial for tumors near but not under the fovea. However, when the tumor is subfoveal or immediately adjacent to the fovea, we most often employ plaque

radiotherapy combined with 3 sessions of extrafoveal TTT in order to adequately treat the tumor but retain useful vision as long as possible. Primary TTT can also be useful for elderly patients, particularly those with diabetes mellitus so that macular edema might be avoided. Photodynamic therapy Photodynamic therapy using verteporfin has been considered for uveal melanoma but little has been published. One report on 4 patients showed tumor regression for 18 months in one patient, but lack of response or continued growth in 3 patients. The ocular oncologist should monitor the uveal scar for tumor regression and complications of therapy. The medical oncologist should survey for metastatic disease. Particular evaluation of the liver, lung, and skin should be made as this malignancy most often metastasizes to these sites. It is recommended that physical examination and liver function testing twice yearly, as well as liver magnetic resonance imaging and chest radiograph annually be performed for monitoring. Surgery There are several available methods for surgical management of posterior uveal melanoma including plaque radiotherapy, charged particle radiotherapy, local resection, enucleation, and exenteration. Radiotherapy Radiotherapy is still the most widely employed treatment for posterior uveal melanoma. The most commonly employed form of radiotherapy is brachytherapy, using a radioactive plaque. Originally, plaque radiotherapy was used for small and medium-sized melanomas located outside the macular region and posterior to the ora serrata. Later, with innovations in radiotherapeutic plan, plaque radiotherapy can be custom designed to treat uveal melanoma at any site within the eye including macular melanoma using a round or notched plaque, juxtapapillary melanoma using a notched plaque, ciliary body melanoma using a round or curvilinear plaque, iris melanoma using a curvilinear plaque, and even extrascleral extension of uveal melanoma. Among the 45 documented recurrences, about one half occurred at the margin, presumably due to treatment planning errors. Recurrence of the tumor was independently related to risk of tumor-related death. Similar to plaque radiotherapy, radiation complications in the eye and adnexa can occur. The COMS conducted three multicenter trials including: In the medium-sized tumor trial 2. Local Resection Local resection of melanomas involving the ciliary body and choroid can be performed using a partial lamellar sclerouvectomy technique. In contrast to radiotherapy it has fewer long-term complications if the initial surgery is successful. However, it does have more potential immediate complications, such as vitreous hemorrhage, retinal detachment and cataract, while radiotherapy is almost uncommonly associated with such immediate complications. However, radiotherapy carries the risk for long-term complications of radiation retinopathy, papillopathy, glaucoma, and cataract. There is no current evidence that local resection of posterior uveal melanoma is different from enucleation or radiotherapy with regard to patient survival. There are fewer complications and better visual results for smaller, more anteriorly located tumors. More complications can be expected when larger post-equatorial tumors are managed in this manner. Enucleation The traditional method of treating uveal melanomas by enucleation was challenged several years ago. Another relative indication for enucleation is melanoma overhanging the optic disc or with optic nerve invasion. Enucleation with a long section of the optic nerve is appropriate in such cases. However, many juxtapapillary melanomas that abut the optic nerve and show no evidence of invasion can be managed by custom-designed notched radioactive plaques rather than enucleation. The "no touch" technique has recently fallen into disuse at most centers because it is cumbersome and its benefits are only theoretical. However a gentle standard technique of enucleation should be employed, without clamping the optic nerve prior to cutting it. There have been recent advances in the types of orbital implants used following enucleation. The hydroxyapatite implant, designed to improve the ocular motility in patients undergoing enucleation, is used widely. However, in the rare instance of massive orbital extension in a blind, uncomfortable eye, primary orbital exenteration is probably justified. In most instances of orbital extension for uveal melanoma, it is not necessary to sacrifice the skin of the eyelid. The eyelid-sparing exenteration provides a better cosmetic appearance. Following radiotherapy there are complications of radiation retinopathy, papillopathy, cataract, glaucoma, scleral necrosis, and pain. Management of Systemic Metastasis Ideally, the best management of uveal melanoma would be to use methods of preventing metastasis in the early stages of the intraocular disease. If the metastasis occurs as a solitary lesion, local resection of the metastatic focus can prolong life. Likewise, the role of chemotherapy and immunotherapy is unproven in the treatment of patients with systemic metastasis from uveal melanomas. There have been reports of tumor

regression after hepatic arterial chemoembolization or immunoembolization. The role of monoclonal antibodies in the detection and management of metastatic uveal melanoma is currently being investigated. Complications Complications of each therapy is detailed in the literature. In general, the main ocular complication following radiotherapy is retinopathy with resultant decreased visual acuity. The treatment of these complications involves laser photocoagulation, anti vascular endothelial growth factor medications, and anti-inflammatory medications. With regards to life prognosis, uveal melanoma prognosis has been shown to be dependent on several clinical factors including tumor location in the ciliary body, large tumor size, diffuse flat configuration, and extraocular extension as well as histopathologic and cytogenetic factors including epithelioid cell type, increased mitotic activity, infiltrating lymphocytes, tumor vascular networks, and chromosomal mutations including monosomy 3 and 8q addition. Clinical factors predictive of metastasis by multivariate analysis included increasing patient age, ciliary body location, increasing tumor diameter, increasing tumor thickness, tumor pigmentation, and subretinal fluid, intraocular hemorrhage, and extraocular extension. The greater the tumor thickness, the greater the risk for enucleation. A Text and Atlas. An Atlas and Textbook. Philadelphia, Lippincott Williams and Wilkins,

**8: Evisceration in Unsuspected Intraocular Tumors | JAMA Ophthalmology | JAMA Network**

*Conservative management is generally advocated whenever possible, but surgical intervention may be justified with unequivocal tumor growth or with extensive disease at initial examination. The management of small choroidal melanomas is controversial, and it is not clear whether treatment of small tumors prevents metastasis.[ 1 ].*

Eye cancer in cow is a very painful and a fatal disorder which start as small nodular growth in the adnexa of eye. Squamous cell carcinoma is regarded as the most frequently diagnosed cancerous tumour in the bovines. The malignant tendencies of this disease make early recognition critical. Human negligence and callous attitude can lead to severe inflicting injuries to the eye which ultimately culminates in eye cancer. I would like to share my experience during my tenure in Animal Rahat, PETA , the largest working animal welfare organization dedicated for working bullocks. We witnessed many animals in a northern Maharashtra village with eye cancer on the right eye. When enquired , it was a shocking finding that the abuse of the eyes by rubbing a combination of pepper and salt to make the animals run during bullock races and sugarcane season had led to high incidence of eye cancer. The cancer growth may start at the medial canthus, lateral canthus or on eyelid. Third eyelid is the most common site of origin of eye cancer. The stage of presentation of the case decides the chances of saving an affected eye. Cases presented in initial stages can be fully recovered and the eyes can be saved. But many cases come in advanced chronic stage with extensive tumor mass eating up the entire globe. Lesions removed in this way are very unlikely to recur. Applying hyperthermia high temperature or cryotherapy freezing to kill the remaining tumor cells in the tissue of the lids but the procedure can not be employed for all cancers. Evisceration surgery involves removal of the contents of the globe while leaving the sclera and optic nerve in place. The cornea can be retained in some evisceration cases. The ultimate goals of these surgeries are to safely and effectively remove the diseased eye or orbital contents using advanced surgical techniques, eliminate the severe underlying ocular pathology Removal of the entire eyeball is indicated when tumors have spread to the extent that the eyeball is blind; the tumor has invaded deeper structures surrounding the globe; or the eyelid is involved to the extent that it cannot be repaired after removal of the tumor. Four point retrobulbar block ensures a painless surgical procedure equally rewarding for the surgeon and for the animal. The 4-point retrobulbar block is technically easier and can be done more rapidly as compared with the Peterson eye block. Introduction of the needle through the conjunctiva should be avoided to reduce the occurrence of ocular contamination. The needle is directed behind the globe using the bony orbit as a guide. Mydriasis indicates a successful block. Other ocular nerve blocks in cattle are Auriculopalpebral Nerve Block: This is a complicated one. After performing a small local skin block over the intended site of puncture, a 3. The cannula is inserted caudal to the junction of the supraorbital process and zygomatic arch and is introduced through the skin. The needle is inserted immediately ventral to the dorsal orbital rim and directed such that the needle impacts into the bone of the orbit. Then the needle is advanced as it is rotated ventrally in a progressive manner such that the needle remains in close proximity to the bone. Successful deposition of lidocaine causes mild proptosis of the globe. Surgical procedure of Exenteration A transpalpebral ablation technique is utilized to remove the eye. A circumferential skin incision is made approximately 1 centimeter from the edges of the eyelids. Using a combination of blunt and sharp dissection, Mayo scissors are used to dissect through the orbicularis oculi muscle, fascia, and subcutaneous tissue surrounding the eye. The interior of the bony orbit is used as a guide. The medial and lateral canthal ligaments are sharply transected to allow access to the caudal aspect of the orbit. As there is a large vessel associated with the medial canthus and optic stalk which need to be ligated after the resection. A vascular clamp can aid in hemostasis while additional excision of remaining orbital tissue is undertaken. In cases where neoplastic infiltration of the bony orbit has occurred, affected areas of ocular periosteum should be thoroughly excised. The skin incision can be closed in a variety of patterns with a nonabsorbable suture such as No. Common patterns include the Ford interlocking, cruciate or simple continuous after sufficiently obliterating the dead space with iodine impregnated gauze rolls. This needs to be changed for 5 days till the cavity dries and heals up. The animal should be kept in a confined area for several days after surgery to allow for appropriate

hemostasis to occur. Daily observation of the surgical site and assessment of general well being is recommended until suture removal. Sutures should be removed in 14 to 21 days to allow for complete healing of the skin.

## 9: Ocular Surface Tumors

*Intraocular eye cancer diagnosis is based on ophthalmic examination, patient history, A/B scan ultrasonography, fluorescein and indocyanine green angiography, and optical coherence tomography. Ocular tumors can be generally divided into the categories that appear below.*

Surgical Management of Intraocular Tumors has been mentioned in several parts of this book, but specific steps have not been illustrated. Techniques illustrated here include fine needle aspiration biopsy FNAB , application of radioactive plaque, and surgical tumor removal by iridectomy, partial lamellar sclerouvectomy, enucleation, and orbital exenteration 1 , 2 , 3 , 4 , 5 , 6 , 7 , 8 , 9 , Laser photocoagulation and cryotherapy are discussed elsewhere 1 , 2 and are illustrated earlier in this book as they apply to specific lesions, Intraocular FNAB is a method of diagnosing selected lesions that defy an accurate diagnosis using less invasive diagnostic modalities 3. It is a rather difficult procedure that requires the cooperation of an experienced ocular oncologist and cytopathologist 3. It should be reserved for selected cases in which an accurate diagnosis will influence the therapeutic choice. It is performed by passing a fine needle into the suspicious tissue and obtaining cells for special preparation and cytologic analysis. In the case of an iris lesion, the needle is passed through the corneoscleral limbus directly into the tumor, using the surgical microscope for better visualization. For posteriorly located lesions, the needle is passed through the pars plana and vitreous and into the tumor, using either indirect ophthalmoscopy or the surgical microscope for guidance. Although false results sometimes occur, it is an accurate diagnostic procedure in most cases 3. Selected iris tumors can be removed by partial iridectomy. Selected ciliary body or choroidal tumors can be removed by techniques of partial lamellar sclerouvectomy. It is used mainly for melanoma, leiomyoma, and tumors of the pigmented or nonpigmented ciliary epithelium 4 , 5. Enucleation is indicated for advanced malignant tumors such as retinoblastoma and uveal melanoma that cannot be successfully managed by other methods 2 , 6 , 7. A gentle technique should be employed in all cases, and the method may vary depending on whether the tumor is a melanoma 2 or a retinoblastoma 6. Orbital exenteration, particularly the eyelid-sparing technique, is reserved for some advanced tumors, particularly uveal melanomas with large degrees of extraocular extension 7 , 8. The techniques employed are subsequently illustrated and discussed in more detail in the references cited. Diagnostic approaches to intraocular tumors. A Text and Atlas. General principles of management. Fine needle aspiration biopsy of suspected intraocular tumors. The Urwick Lecture. Surgical approach to lamellar sclerouvectomy for posterior uveal melanomas. The Schoenberg Lecture. Partial lamellar sclerouvectomy for ciliary body and choroidal tumors. Enucleation technique for children with retinoblastoma. J Pediatr Ophthalmol Strabismus ; Enucleation for uveal melanoma. Principles and Practice of Ophthalmology, 3rd ed. Orbital exenteration with eyelid sparing: Massive orbital extension of posterior uveal melanoma. J Ophthalmic Plast Reconstr Surg ;7: Instrumentation and Technique The instrumentation, techniques, limitations, complications, and results for FNAB of suspected intraocular tumors and inflammations are reported in more detail in the references cited and are only illustrated briefly here. Fine needle aspiration biopsy of iris tumors in consecutive cases. Instruments used for intraocular fine needle aspiration biopsy. Translimbal, transaqueous approach for iris lesions, front view. The surgical microscope is generally used for this technique. Translimbal, transaqueous approach for iris lesions, side view. Trans-pars plana, transvitreal approach for ciliary body and choroidal lesions. Indirect ophthalmoscopy is usually used to visualize the lesion and facilitate needle guidance. The technique requires considerable experience. Method used for a choroidal mass overlying a bullous retinal detachment. An equatorial sclerotomy is performed, the choroid is cauterized, and the needle is passed obliquely through the subretinal space. This approach avoids inducing a retinal hole. Trans-pars plana approach for fine needle aspiration biopsy of free cells in the vitreous cavity. This technique is most appropriate for diagnosing vitreous lymphoma or differentiating conditions such as vitreous melanoma cells from blood cells. Standard vitrectomy techniques can also be used to make the diagnosis in such cases. Superior iris mass in a 6-year-old boy. Fine needle aspiration biopsy was performed. Cytopathology, showing

histiocytes and a giant cell, findings compatible with juvenile xanthogranuloma. Two peripheral iris masses in a year-old man, one superotemporally and one superonasally. Cytopathology revealed malignant cells compatible with lung cancer. Subsequent systemic evaluation revealed an occult lung cancer. Pigmented iris lesion with tumor seeding on the iris surface in a year-old man. Cytopathology of fine needle aspiration biopsy on the lesion shown in Figure A radioactive plaque was applied. Mass inferior to the optic disc in a year-old woman. The lesion appeared to have a pigmented base, and melanoma was suspected. However, the patient had a history of breast cancer, and metastasis was considered to be a possibility. Cytopathology revealed carcinoma, compatible with metastatic breast cancer. Syringe and connector tubing used for fine needle aspiration biopsy. Calipers set at 4 mm to ensure entrance through the pars plana by the gauge needle. Needle being passed through the conjunctiva, sclera, and pars plana into the vitreous cavity. The needle is then passed through the vitreous into the tumor with indirect ophthalmoscopy guidance or surgical microscope guidance. After tumor tissue is aspirated into the needle bore, balanced salt solution is aspirated through the needle into the syringe. The solution is then submitted to cytopathology laboratory for studies. Some prefer to smear the aspirated material on a slide, which is stained for cytopathologic study. Cytopathology of an aspiration of the lesion shown in Figure

Plaque Design and Application Figure Standard round, mm iodine plaque. The dummy plaque is to the left and the shielded side of the active plaque is to the right. Opposite side of the active shielded plaque, showing the iodine seeds. The side shown here is directed toward the sclera at the time of plaque application. Standard round plaque being positioned at the time of surgery. Notched plaque for the treatment of juxtapapillary tumors. Custom-designed plaque for treating ciliary body tumors. The plaque shown in Figure It can be used under special circumstances for circumscribed choroidal hemangioma, retinal vascular tumors, and perhaps other intraocular lesions. The use of radioactive plaques for these tumors is illustrated under the specific chapters in this book. Only gold members can continue reading. Log In or Register to continue Share this:

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