Symposium

Vascular tumors of the choroid and retina

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Vascular tumors of the retina and choroid can be seen occasionally. In the following article, the key clinical and diagnostic features of the major retinal and choroidal vascular tumors, their systemic associations, and the literature pertaining to the most currently available treatment strategies are reviewed.

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Vascular tumors of the choroid such as the solitary and diffuse choroidal hemangioma and those of retina, such as the capillary, cavernous, racemose hemangioma and vasoproliferative tumor (VPTR) of the retina are covered in this chapter.

Choroidal Hemangioma

Choroidal hemangioma is a benign vascular tumor and is of two types. The circumscribed solitary tumor which has no extraocular associations while the diffuse type can be associated with facial naevus flammeus or variations of Sturge-Weber syndrome.[1] Cavernous hemangioma of the choroid is the most common vascular tumor of the uveal tract.

Circumscribed Choroidal Hemangioma

Circumscribed choroidal hemangiomas (CCHs) are relatively rare hamartomatous vascular tumors generally diagnosed in patients between 20 and 40 years of age.[1] The tumor is almost always unifocal and unilateral with bilateral cases being extremely rare.[2]

Clinical features

Circumscribed choroidal hemangioma appears as a discrete, smooth, homogenous surface, round or oval, orange reddish mass located in the posterior pole.[3-5] The tumor is usually located posterior to the equator with most in the macular or peripapillary region.[3] The tumor size may range from 3 to 18 mm (mean 7 mm) in diameter and 1–7 mm in elevation (mean 3 mm).[4]

Subfoveal tumors produce hyperopic shift secondary to the anterior displacement of the retina.[1] Patients with parafoveal tumors may remain asymptomatic unto adulthood, when they present with either gradual or sudden onset of blurred vision secondary to exudative retinal detachment (RD).[1] The retina over a CCH often develops severe secondary cystoid retinal degeneration.[1] Occasionally, a total RD can occur and lead to neovascular glaucoma.

Ancillary testing

Fluorescein angiography

Circumscribed choroidal hemangioma show some characteristic features on fundus fluorescein angiography (FFA) but are not pathognomic.[1-4] The large choroidal vessels of the tumor show irregular linear hyperfluorescence in the prearterial or the early arterial phases.[4] In the arterial and venous phase there is progressive profuse leakage from pinpoint foci of hyperfluorescence.[1] In the late phases, intraretinal hyperfluorescence is seen due to the diffuse leakage of dye into the cystic spaces in the retina overlying the tumor.

Indocyanine green angiography[5]

Hyperintense indocyanine green (ICG) fluorescence is seen at an average of 1.2 min after injection. A lacy diffuse fluorescent pattern is seen in the early phases, leading to hypofluorescence of the tumor with dye wash out in the late phase. A rim of hyperfluorescence is seen surrounding the tumor in the late frames.

Autofluorescence[6]

Choroidal hemangioma shows little intrinsic autofluorescence. Overlying lipofuscin and fresh subretinal fluid (SRF) show hyperautofluorescence and retinal pigment epithelial (RPE) hyperplasia and atrophy show hypoautofluorescence.

Ultrasonography

Circumscribed choroidal hemangioma show high anterior spike and high internal reflectivity on A scan ultrasonography with no significant shadowing and acoustic solidity on B scan. Occasionally, high reflective echoes can be noticed over tumor

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surface due to dense fibrous or osseous metaplasia of the overlying RPE.[1,3,4]

Magnetic resonance imaging

On magnetic resonance imaging (MRI), CCH is relatively hyperintense with respect to the vitreous in T1-weighted images and isointense to the vitreous in T2-weighted images, in contrast to a melanoma.[4,7] The tumor shows a marked enhancement on gadolinium administration.

Pathology

Histological classification includes three types.[1] The capillary type which is rare and is composed of small blood vessels with inconspicuous endothelial cells separated by loose connective tissue. The cavernous type which is more common and consists of large, thin-walled blood-filled vascular channels lined by a flat endothelium and separated by thin intervascular septa.[1] A mixture of both types can be seen in many tumors.

Extensive cystic changes are generally present in the outer retinal layers overlying the tumor and may coalesce to form retinoschisis. The RPE overlying the tumor may show fibrous or osseous metaplasia. The pigmented rim seen clinically is caused by irregularly compressed choroidal melanocytes at the margin of the hemangioma.[1]

Differential diagnosis

Amelanotic choroidal melanoma

A typical CCH has a red-orange color, which is never seen with a choroidal melanoma. The characteristic findings of acoustic hollowness and choroidal excavation seen in choroidal melanomas on ultrasonography are almost never seen with a CCH.

Choroidal metastasis

Choroidal metastasis usually has a dull or creamy yellow and often multifocal and bilateral in comparison to unilateral and unifocal with red-orange color of a CCH.[8]

Posterior scleritis[9]

Posterior scleritis can sometimes resemble a CCH but is usually associated with inflammatory signs and choroidal folds.

Management

The management of choroidal hemangioma varies with the severity of symptoms and the presence or absence of exudative RD.[4,10] In asymptomatic cases, no treatment is necessary as CCH rarely increases in size and remains quiescent for months or years with periodic review.[1]

Treatment is usually considered in cases with visual loss. The various treatment options are photodynamic therapy (PDT), photocoagulation[4,10,11] external beam irradiation, (external beam radiation therapy)[12,13] proton beam therapy, brachytherapy,[14] and transpupillary thermotherapy (TTT).[15]

Photocoagulation

Green or rarely red wavelength laser photocoagulation is used to create a chorioretinal adhesion and resolve the SRF.[4,15] The treatment should be intense enough to produce a slight white reaction on the surface of the tumor. In cases of extensive RD surgical drainage of SRF can be carried out to allow photocoagulation.[1]

In TTT, infrared laser is employed to raise the internal tumor temperature above 45°C but below 60°C.[15] The chorioretinal scar observed after treatment is less pronounced in comparison to photocoagulation. ICG can be used to augment the effect of TTT.[16] TTT can result in complications such as cystoid macular edema (CME), preretinal fibrosis, and retinal vascular occlusion making TTT unsuitable for subfoveal and peripapillary tumors.[17]

Photodynamic therapy [Fig. 1a-c]

Photodynamic therapy offers site-specific tumor destruction while sparing overlying retina and retinal vasculature.[18] Selective occlusion of choroidal neovascularization can be achieved while the neurosensory retinal layers and Bruch membrane are almost unaffected, leaving retinal function intact.[19] Currently, PDT is the preferred treatment option for managing choroidal hemangioma, particularly those involving the macula.[1,20,21] Visual improvement or stabilization has been reported in 73–100% of treated patients most often with a single treatment.[22,23]

Various investigators have employed differing treatment protocols including verteporfin injection parameters (bolus versus injection over 10 min), total number of treatment sessions (1–5 sessions), laser power settings (50–100 J/cm^2), duration of exposure (83–186 s), and number of laser spots (one or more). It is preferable to wait for 4–6 months after PDT for resolution of SRF, before repeating the treatment as repeat PDT may result in RPE atrophy and vision loss.

Proton beam therapy

As charged particles have a highly localized and uniform dose distribution proton beam therapy was one of the options to treat macular and juxtapapillary CCH prior to PDT.[24] Contradictory report regarding the usage of proton beam therapy in such conditions have been published with Zografos *et al*.[24] have advocated proton beam as optimal for treating these tumors in this critical region while Lee and Hungerford have reported that proton therapy has been disappointing in preventing radiation optic neuropathy and maculopathy and offers little advantage over external irradiation.[25]

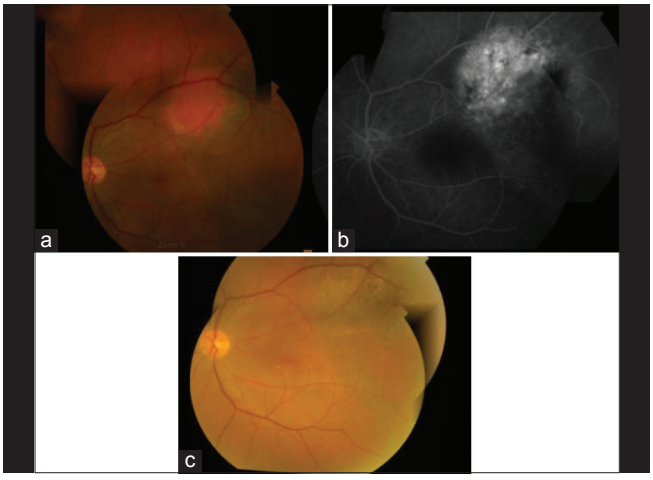

Figure 1: (a) Solitary discrete, smooth, oval, orange reddish mass in the superior temporal quadrant confirming circumscribed choroidal haemangioma. (b) Shows early hyperflourescence with intense leakage. The inferior extent of the leakage is well-appreciated in the form of retinal pigment epithelial tracts. (c) The same patient after full fluence photodynamic therapy showing complete regression of the tumor

Brachytherapy

Brachytherapy can be employed in circumscribed hemangioma in certain special circumstances which include, hemangioma with extensive SRF where PDT would not be feasible, juxtapapillary tumors not responding to PDT or TTT. Low-dose radiation delivering 20 Gy to the tumor apex is adequate. Visual loss due to radiation retinopathy and optic neuropathy may occur in up to 30% of patients despite the low dosage utilization.[26] Tumor regression, resorption of SRF and regression of iris neovascularization have been reported with I-125 and palladium-103 brachytherapy.[26-28]

External beam radiation therapy

Though more reserved in treating diffuse choroidal hemangioma, Kong *et al*. have reported three patients treated with a maximal dose of 10 Gy with good anatomical and functional outcome with no side effects noted in the follow-up period of 18–36 months.[29]

Single or multiple antivascular endothelial growth factor (VEGF) bevacizumab intravitreal injections with or without PDT or TTT has been used to resolve SRF associated CCH.[30,31]

Oral propranolol has recently found efficacious to partially or completely resolve SRF associated with CCH.[32,33] Larger, comparative prospective studies are likely to define the role of anti-VEGF injections and oral propranolol to treat choroidal hemangioma in the future.

Visual and anatomical prognosis

Shields *et al*. have noted complete resolution of SRF occurred more often in patients with shorter duration of symptoms and inferior quadrant location of the tumor. Poor initial visual acuity, failed prior laser photocoagulation, and delay in referral are predictors of poor final visual acuity.[34]

Diffuse Choroidal Hemangioma

Diffuse choroidal hemangioma is a benign vascular tumor that has poorly defined borders and extends over a broad area of the posterior choroid.[1] It is characteristically associated with cutaneous, ocular or central nervous system findings, sometimes as part of the Sturge-Weber syndrome. The precise incidence of diffuse choroidal hemangioma in patients with Sturge-Weber syndrome is not known. A series of 35 patients with this syndrome found an incidence of 31% of diffuse choroidal hemangioma and a 100% incidence of facial naevus flammeus.[35]

Clinical features

Diffuse choroidal hemangioma is usually diagnosed at a young age, either because the associated facial hemangioma prompts a fundus examination or due to visual impairment caused by hyperopic amblyopia or from a secondary RD.[1] The pupil shows a brilliant red reflex (tomato catsup fundus) in the involved eye in contrast to the normal reflex in the opposite pupil.[36]

Diffuse choroidal hemangioma on ophthalmoscopy reveals a diffuse red-orange thickening of the posterior choroid, which is mainly seen in the macular area. Cystoid degeneration in the overlying retina over the tumor surface with RPE disruption commonly occurs. In comparison to CCH, the diffuse choroidal hemangioma is frequently large and often extends anterior to the equator. Ipsilateral congenital glaucoma can develop, particularly when the upper lid is involved by the naevus flammeus. Asymmetry of the optic disc can be documented with enlargement of the optic cup.[37] Vascular (increased episcleral venous pressure), mechanical, and developmental causes offer the most plausible mechanisms to explain the pathogenesis of glaucoma in encephalotrigeminal angiomatosis.[38] The exudative nature can be precipitated by prostaglandin analogs used to treat the accompanying glaucoma in patients with associated Sturge-Weber syndrome.[39]

Diagnosis

Ultrasonography demonstrates a markedly thickened choroid with medium to high internal reflectivity with an overlying RD. FFA reveals diffuse leakage similar to CCH but with more widespread involvement. MRI features are similar to CCH.

Pathology

Diffuse choroidal hemangioma is characterized by an intermixed proliferation of small and large blood vessels and is usually classified as a mixed hemangioma. Fibrous transformation of the proliferated RPE is observed in over 50% of diffuse choroidal hemangiomas in Sturge-Weber syndrome.

Management

Diffuse choroidal hemangioma is a difficult condition to manage. Hyperopic refractive error can be addressed with refraction, corrective lenses and amblyopia therapy.[1] With extensive secondary RD, drainage of SRF and or scleral buckling with pars plana vitrectomy and injection of gas and endolaser may be recommended.[1]

Glaucoma management is also complicated as serous or hemorrhagic choroidal detachments are a common with glaucoma filtering surgery in patients with diffuse choroidal hemangioma.

External beam radiation therapy in the dose of 1250–2000 cGy in divided fractions has been utilized for treatment of diffuse choroidal hemangiomas.[40] Radiation leads to resolution of the RD in most cases and control of glaucoma in some cases. Plaque brachytherapy can also be employed to treat eyes with the plaque being centered on the thickest part of the tumor.[41]

Photodynamic therapy is the preferred option to treat diffuse choroidal hemangioma wherein the SRF would allow therapy. A single spot over the thickest part of the tumor or multiple nonoverlapping spots of single or multiple session PDT has resulted in resolution of exudative RD and regression of the tumor.[42]

Propranolol, a nonselective β blocker in the dosage of 2 mg/kg/day has been recently noted to cause accelerated absorption of the exudative RD in some cases, the mechanism being unclear. Propranolol interferes with endothelial cells, vascular tone, angiogenesis, and apoptosis and may thus lead on to the resolution of the RD.[43] A single intravitreal bevacizumab injection has resulted in sustained resolution of exudative RD in Sturge-Weber syndrome over 20 months in a single case.[44]

We have treated a diffuse choroidal hemangioma with bullous RD resistant to systemic propranolol and brachytherapy, with drainage of SRF and intraoperative TTT, resulting in resolution of the RD, regression of the tumor, and control of associated glaucoma (unpublished data: Fig. 2a-c).

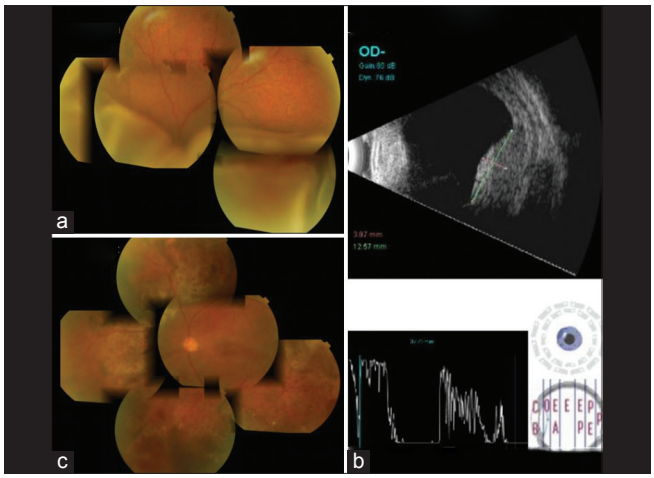

Figure 2: (a) A patient with diffuse orange reddish mass occupying the entire posterior pole s/o diffuse choroidal hemangioma. (b) Ultrasound A and B scan showing high surface with high to medium internal reflectivity and exudative retinal detachment. (c) Complete resolution of exudative retinal detachment with regression of the tumor mass after subretinal fluid drainage and transpupillary thermotherapy

Vascular Tumors of the Retina

Capillary hemangioma of the retina

Capillary hemangioma of the retina may occur as an isolated lesion within the retina or as a part of phakomatosis with central nervous system and systemic tumors when it is called von Hippel-Lindau (VHL) syndrome.[45] It is genetically transmitted in an Alzheimer's disease (AD) model with incomplete penetrance and variable expressivity. VHL syndrome is linked to a defect in a short arm of chromosome 3 (3p 25-26).[46] The mean age of diagnosis is 18 years for patients with VHL and 36 years for non-VHL. Development of the associated systemic tumors occurs early in life and very rarely after 40 years.[47] In a patient with solitary hemangioma, the risk of developing VHL is 45% if the patient is <10 years of age and decreases to 1% if the age at diagnosis is >60 years.[48] Central nervous system hemangiomas occur in more than 50% of patients with VHL, and they are diagnosed at a younger age than in sporadic cases. The most commonly involved sites include the cerebellum (75%) and the spinal cord (15%).[49] Systemic manifestations of VHL include pheochromocytoma, hypernephroma, pancreatic cysts, endolymphatic sac tumor, and several other tumors and cysts. Renal cell carcinoma is the leading cause of mortality in patients with VHL, occurring in 5% of patients by age 30 and 40% by 60 years of age.[48] Pheochromocytomas are observed in <25% of patients with VHL. When associated with VHL, these tumors tend to be multiple and bilateral.[50] Capillary hemangioma of the retina can also be associated with other systemic conditions like Marshall-Stickler syndrome.[51]

Clinical findings[1]

The ocular lesions are usually diagnosed between 10 and 30 years of age. The early angioma appears as a yellow spot between a dilated tortuous feeding arteriole and a draining venule. Vision loss occurs due to exudation at the macular region, (which is usually due to the steal phenomenon or subretinal migration of lipid from the periphery) or secondary tractional/rhegmatogenous RD (due to development of gliotic tissue) involving the macula. An appearance of dilated pair of vessels in the posterior pole with macular exudates should prompt one to examine the periphery for a capillary angioma. The most common retinal location of these tumors is in the superotemporal quadrant (42%) and the midperipheral (58%).[1] Multiple or bilateral angiomas indicate the presence of VHL tumor and screening for central nervous system or systemic disease should be done.

Some authors believe that two forms of the disease exist – the exudative form and the vitreoretinal form.[1] In the vitreoretinal form, epimacular membranes form which cause macular traction detachment and decreased vision. Traction on the angiomas can lead to "free floating" angiomas in the vitreous, vitreous hemorrhage, and a combined traction rhegmatogenous RD can also occur.

Angioma of the optic nerve head may occur. This differs from the retinal angioma in that no feeder arteriole or a draining venule is seen. The orange-red lesion is ill-defined, involves an eccentric part of the optic disc with exudation involving the peripapillary region and simulates papilledema.

Without treatment, most eyes progress to total RD, neovascular glaucoma, and a painful blind eye.

Differential diagnosis

The retinal angioma may be mistaken for Coat's disease (no paired dilated vessels in Coat's disease), racemose angioma (no tumor between the arteriole and venule in racemose angioma), intraretinal macroaneurysm (occurs along a arteriole without a draining venule), retinal cavernous hemangioma (no feeder vessels; multiple sac like aneurismal dilatations), familial exudative vitreoretinopathy, and nematode endophthalmitis.

The disc angioma may simulate papilledema, optic neuritis, peripapillary choroidal neovascular membrane, and optic disc granuloma.

Pathology[52]

The lesions are composed of benign proliferation of spindle and endothelial cells, pericytes, small blood vessels, and clear stromal cells. Currently, retinal hemangioblastoma appears to be the preferred terminology as they mimic the cerebellar hemangioblastoma very closely.[49]

Diagnosis

The diagnosis can be made most often on indirect ophthalmoscopy with the classic picture of an orange tumor with a feeding arteriole, draining venule associated with macular exudates. An FFA may, however be necessary to delineate early lesions that are not visible on clinical examination. These lesions fill early phases of the angiogram and leak profusely later. The paired vessels are well-delineated as well.

Treatment

The choice of treatment is determined by the size, location, and associated findings of SRF, retinal traction, and the visual potential of the eye.[53]

It is also been proven that early treatment of retinal capillary angiomas leads to better visual results.[54] Careful observation in a reliable patient can be recommended if the retinal capillary hemangioma is very small (up to 500 microns), and positioned

in the juxtapapillary area as they tend to remain stable as well as those not associated with exudation or SRF, and is not visually threatening.[55]

Early angiomas not at the juxtapapillary areas without RD are treated with laser photocoagulation. It is preferable to use green, yellow or blue-green wavelength to treat these lesions, as red or infrared lasers may not be absorbed well. Lesions < 2 mm are treated with a direct photocoagulation [Fig. 1a]. For lesions 3–5 mm, it is preferable to try occlusion of the feeder vessels – the arteriole in the first session and the venule later. The angioma as such can be treated in subsequent sessions. Hemorrhage from the tumor and increase in secondary RD may occur as complications.

For tumors larger than 5 mm, it is preferable to use triple freeze-thaw cryotherapy. Increase in exudative RD and hemorrhage can occur as a complication of cryotherapy and settles with time.

If the tumors that are associated with a bullous RD, drainage of the fluid, cryotherapy, and scleral buckling may be necessary. Advanced vitreoretinal form of the disease may need vitrectomy [Fig. 3b and c] to relieve tractional or rhegmatogenous RD.[56]

Transpupillary thermotherapy and plaque brachytherapy have been employed in the management of retinal and disc angiomas.[57,58]

Proton beam irradiation and external beam radiotherapy have been used as salvage treatment when other treatments have failed.[59]

Other methods of limited use include PDT, but when used to treat peripapillary angioma regresses the tumor and resolves macular detachment, however may be associated with risk of iatrogenic retinal vascular occlusion and optic disc ischemia.[60] Transretinal feeder vessel ligation combined with vitrectomy and photocoagulation has also been reported in the literature.[61]

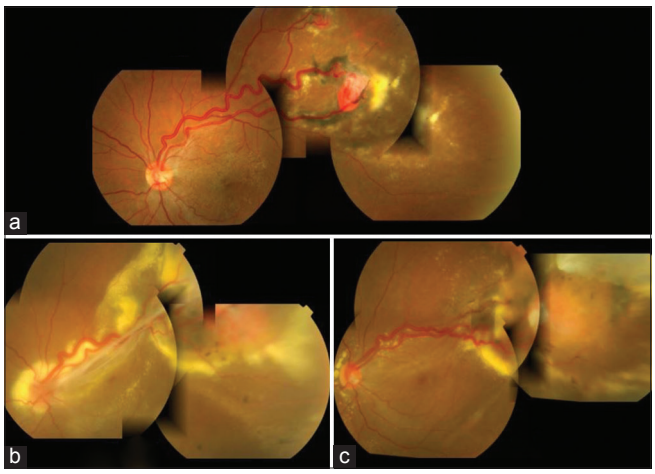

Figure 3: (a) A patient a retinal hemangioblastoma with well-seen dilated feeder vessel and accompanying draining venule. Laser photocoagulation was done to the mass lesion and the feeder vessel. (b) 2 years after treatment, the patient developed traction bands from the disc to the lesion and causing vitreomacular traction and subretinal exudates. (c) The patient underwent pars plana vitrectomy and membrane peeling. The macula is settled back with a resolution of the subretinal exudates

Bevacizumab and ranibizumab have shown some beneficial effect on subretinal exudation and CME associated with retinal angioma. Multiple injections are required, and treatment is often combined with PDT and intravitreal or sub-tenon triamcinolone. The efficacy appears to be limited to the exudative component and did not bear any significance in the size of the lesions or the occurrence of newer ones.[62]

Systemic features[63]

Von Hippel-Lindau syndrome does not have major cutaneous features, with occasional occurrence of nevi, or café' au lair spots. Cerebellar hemangioblastomas is the classic central nervous system lesion and usually causes symptoms in the 4th decade of life. Treatment is in the form of surgical resection is possible.

Periodic ocular, neurological, and systemic evaluation with MRI of the brain and spine once in 2–3 years (every 3–5 years in patients >51 years), urinary catecholamines and abdominal ultrasound/computerized tomography every year of patients and screening of family members at risk is very important.

Cavernous hemangioma of the retina

Benign vascular tumor most commonly diagnosed in children or young adults and may be associated with intracranial hemorrhage resulting in stroke, cranial nerve palsies, and their related signs and symptoms.[64] Although retinal cavernous hemangioma is usually considered to be the only ocular manifestation of this AD condition, other rarely reported ocular features include choroidal hemangioma[65] and ocular melanocytosis.[66]

Peripheral lesions are asymptomatic, macular lesions result in loss of vision.[1] Ophthalmoscopically, the lesions appear as a cluster of dark red saccules (blood-filled saccules) with associated fibroglial proliferation within the inner retinal layers or on the surface of the optic disc.[1] The lesions are variable in size and location, and frequently follow the course of a major retinal vein. They are usually nonprogressive or may enlarge and cause vitreous hemorrhage. Epiretinal membranes and fibroglial proliferation may, however, result in foveal ectopia and visual loss.[67] Massive growth of the tumor up to the iris root with concomitant vitreous hemorrhage and hyphema have been reported rarely.[68]

Diagnosis

The diagnosis is evident on fundus examination, but the FFA is quite characteristic. These lesions have a slow blood flow which leads to the separation of the plasma from the blood cells, which settle down within the saccule and appear as "fluorescein cap."

No treatment is required for this lesion. If vitreous hemorrhage occurs, vitrectomy, cryotherapy, photocoagulation, low energy plaque may be used to treat these tumors.[1]

Racemose hemangioma

Racemose hemangioma is more of a vascular malformation than a tumor and if part of phakomatosis, is called the Wyburn-Mason syndrome. Wyburn-Mason syndrome (also known as Bonnet-Dechaumme-Blanc syndrome)[69] was first described in 1943 when the association was made between racemose hemangiomatosis of the retina and arteriovenous malformations of the brain.[70] No definite hereditary pattern has been noted. The arteriovenous communications in Wyburn-Mason syndrome have been classified into three types.[71]

The clinical appearance is characteristic, and a fluorescein angiogram may show rapid filling of the arteriovenous communication without dye leakage.

Most lesions are stationary and do not need treatment. However, the visual prognosis is poor. Spontaneous cerebral or subarachnoid hemorrhages may lead to neurological symptoms (hemiplegia, hemiparesis, cerebellar dysfunction, parinaud's syndrome, seizures). Facial (50% of patients, ipsilateral to the side of racemose hemangioma) and orbital hemangiomas, rarely hemangiomas involving the lung, spinal cord may co-exist.[72] Wyburn-Mason syndrome commonly does not have cutaneous involvement. Hemangioma involving the bones of the skull, maxilla, and mandible may lead to massive bleeding during dental extraction. Orbital vascular malformations may cause pulsating proptosis. Ptosis or a partial ophthalmoplegia due to III nerve involvement by intracranial tumor may occur. Intraocular racemose hemangioma communicating with intramuscular facial hemangioma has been reported.[73]

Vasoproliferative tumor of the retina

Retinal VPTRs are very rare and only recently recognized as a distinct clinical entity in the early 1980s.[74] VPTR is very rare solitary tumors and usually occur without any antecedent cause, or may occur in eyes with prior uveitis, retinitis pigmentosa, Coat's disease, FEVR, and toxoplasma scars.[75,76]

They appear as solitary pinkish-yellow raised vascular mass in the preequatorial retina, classically located inferiorly with minimally dilated feeder vessels, associated with intraretinal and extensive subretinal exudation and hemorrhage, secondary RD, premacular fibrosis, tractional RD, RPE hyperplasia, macular edema, and vitreous hemorrhage [Fig. 4a-c]. Some patients may have multiple or diffuse tumors. The pathogenesis of these lesions is unclear.

Fluorescein angiography shows early filing and late leakage and dilated feeder vessels [Fig. 4c and d].

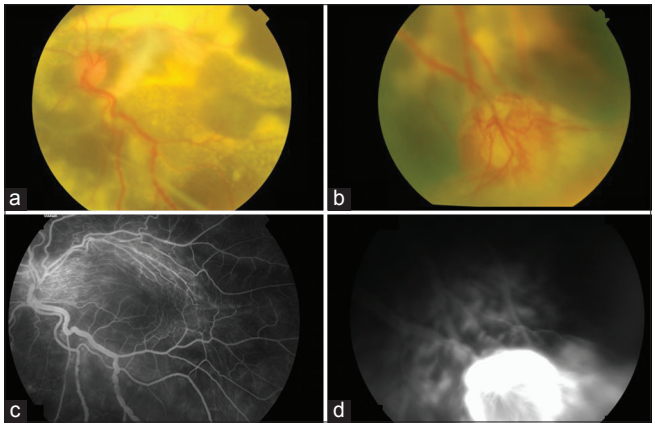

Figure 4: (a) A patient with massive subretnal exudates with shallow exudative retinal detachment. (b) Careful peripheral examination showed a yellowish mass lesion with intense vascular supply s/o vasoproliferative tumor. (c and d) Fundus fluorescein angiography of the same patient showing dragging of the vessels at the posterior pole with very early hyper-fluorescence and intense leakage. This patient was treated with intravitreal tricort in the dosage of 4 mg/0.1 ml with triple freeze-thaw

Treatment is indicated if progressive exudation results in vision loss while some believe in treating prior to the occurrence of symptoms.[75-79]

Multiple repeated sessions of triple freeze-thaw cryotherapy are often the preferred treatment option for VPTR.[75] Both I-125 and ruthenium-106 brachytherapy have been used to treat VPTRs.[80,81] I-125 brachytherapy has shown tumor regression in 97%, resolution of exudative RD in 65%, and improvement of visual acuity in 73% in a case series.[80]

Photocoagulation and PDT have been used in a limited number of cases with some success.[75,82,83] More recently, case reports of successful usage of bevacizumab in the treatment of VPTR can be found in the literature.[84]

Vitrectomy is generally reserved for cases of VPTR resulting in tractional RD and macular pucker.[76] Advanced cases may eventually develop neovascular glaucoma requiring the enucleation.[75]

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