Choroidal and Retinal Tumors in Adults: Vascular Tumors of the Retina

Part one of a four-part series on guidelines for diagnosing and treating ocular neoplasms.

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Cataracts normally occur in the later decades of life, which is also at a time when there is higher incidence of ocular tumors. Before a patient undergoes cataract surgery, it is important that surgeons perform a complete eye examination that includes careful funduscopy. The surgeon must be able to distinguish ocular tumors from other lesions and, should a lesion be identified, send patients for proper evaluation before performing cataract surgery.

There are masqueraders of tumors, and there are situations where metastases mimic other diseases. Metastases to the retina are rare. Vitreoretinal lymphoma is also a rare occurrence that should be considered in all cases of intermediate or posterior uveitis. Masqueraders for retinal tumors include inflammatory lesions such as granulomas. Choroidal tumors, choroidal hemorrhages, choroidal effusions, posterior scleritis, and choroidal granulomas must be considered in the differential diagnosis.

In this first article of a four-part series on retinal and choroidal tumors, we describe the clinical features, diagnosis, and treatment for four types of vascular tumors of the retina.

RETINAL HEMANGIOBLASTOMA

Retinal hemangioblastomas, also incorrectly termed capillary hemangiomas, occur in the retina most often in association with von Hippel-Lindau (VHL) syndrome, an inherited multisystem disorder characterized by abnormal growth of blood vessels. Evaluation for other clinical findings associated with VHL, including renal cell carcinoma, central nervous system hemangioblastomas, endolymphatic sac cysts, and pheochromocytoma should be performed. Family members should be evaluated, and medical genetics consultation should be considered.

Clinical features. Red, circumscribed, round retinal lesions with dilated feeding and draining vessels are the primary clinical features of retinal hemangioblastoma. Capillary hemangioblastomas can be found in the juxtapapillary area or elsewhere in the retina. They can be exophytic, in which case the reddish lesion is difficult to visualize. These are most often seen near the disc. These tumors can also be endophytic, in which case the reddish lesion is easily seen. Exudation can dramatically affect visual acuity when the macula is involved (Figure 1).

Diagnosis. Fluorescein angiography (FA) is useful for diagnosis because these kind of lesions have a marked degree of vascularity. The dilated feeding and draining vessels are readily seen, and there is marked hyperfluorescence from fluorescein leakage. If the round mass is large enough, it can be detected on ultrasound B-scan. A-scans show medium to high internal reflectivity. Subretinal fluid can be diagnosed with optical coherence tomography (OCT) or B-scan.

Differential diagnosis. The differential diagnosis of these
lesions includes retinal vasoproliferative tumors, in which the feeding and draining vessels tend not to be dilated, as opposed to retinal hemangioblastomas, astrocytomas, and inflammatory diseases including granulomas and vascular diseases. The differential diagnosis for juxtapapillary hemangioblastomas includes papilledema, papillary neovascularization, disc metastases, and optic nerve glioma.

**Treatment.** The type of treatment depends on the dimensions of the lesion, the presence of subretinal fluid, and visual function. The prognosis is related to these characteristics. Options include observation, laser photocoagulation, cryotherapy, radiotherapy, and antiangiogenic agents. Results with radiotherapy and antiangiogenic agents have been disappointing. Only those with extensive experience in following these cases should observe, because hemangioblastomas tend to grow without treatment.

**CAVERNOUS HEMANGIOMA OF THE RETINA**

**Clinical features.** Cavernous hemangioma is a rare congenital lesion that appears commonly as a solitary vascular lesion of limited size. It affects both men and women and occurs in all ethnicities. Most affected patients have a single lesion in one eye and no evidence of a multisystem syndrome. Intracranial cavernous hemangiomas and angiomatous lesions of the skin can also be seen. These patients should be questioned as to whether they have had central nervous system bleeding or headaches and whether family members have had similar problems. Magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) of the brain should be performed in these patients. The KRIT1/CCM1 gene has been associated with cavernous hemangiomas. Most cases are asymptomatic, but vascular hamartoma can affect visual function by causing small vitreous hemorrhages.

**Diagnosis.** Ophthalmoscopy reveals a characteristic multilobulated dark red lesion in the inner layers of the retina or on the optic nerve with a grape-cluster type configuration. On FA, this lesion shows low flow with a delayed filling during the venous phase, and a layering of fluorescence is seen within each separate lobule. Ophthalmoscopic features are generally sufficient to make the diagnosis.

**Treatment.** If visual acuity is not affected, observation is the best option. Laser photocoagulation has been used when visual function is diminished.

**RACEMOSE HEMANGIOMATOSIS OF THE RETINA**

**Clinical features.** Some systemic arteriovenous malformations (AVMs) are found principally in the brain and less frequently in the maxilla and pterygopalatine fossa, kidneys, or skin. There are three forms of racemose hemangioma, which is also known as Wyburn-Mason syndrome. The first, Archer type 3, occurs between the central retinal artery and central retinal vein and has a high association with systemic disease. The second, Archer type 2, are branch artery and vein malformations that rarely have systemic associations. The third, Archer type 1, involve small-vessel AVMs and do not have systemic associations.

**Diagnosis.** The characteristic finding for type 1 AVMs is tortuosity and dilation of the retinal vessels from the optic nerve to the retinal periphery on funduscopic examination. In FA, rapid transit right into the retinal vein via the AVMs can be observed. Ultrasonography and OCT can also help confirm the diagnosis, and an MRI should be done to exclude intracranial malformations.

**Treatment.** Most patients with large AVMs have decreased visual acuity. An effective treatment has not been established for type 3 lesions.

**RETINAL VASOPROLIFERATIVE TUMOR**

These tumors are rare and can be primary or secondary to multiple associated ocular conditions.

**Diagnosis.** Fundus examination shows a solitary pink-to-yellow vascular mass usually in the inferior retina. Subretinal exudation and hard exudates may be associated, but dilated feeding and draining vessels are not seen as commonly as they are with retinal hemangioblastomas (Figure 2). Evaluation for other ocular diseases associated with vasoproliferative tumors, including pars planitis and retinitis pigmentosa, should be conducted. FA should be performed using a wide-field technique to look for clues to the cause of the vasoproliferative tumor. A-scan shows high internal reflectivity in vasoproliferative tumors that are big enough to be visualized by ultrasonography. OCT can be useful to follow subretinal fluid.

**Differential diagnosis.** Differential diagnosis of retinal
vasoproliferative tumors includes retinal hemangioblastoma, choroidal metastases, and choroidal melanoma.

Treatment. The treatment plan should be based on visual function and subretinal exudation. Options include observation, cryotherapy, brachytherapy, laser photocoagulation, photodynamic therapy, and intravitreous injection of antiangiogenic agents. Usually laser photocoagulation is the best treatment in these cases because the tumors tend to be much smaller than the larger tumors that can occur with VHL syndrome.

CONCLUSION

Subsequent installments in this series will examine choroidal metastases, choroidal melanocytic lesions, intraocular lymphomas, vascular tumors of the choroid, and other tumors.

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• Retinal hemangioblastomas are red, circumscribed, round retinal lesions with dilated feeding and draining vessels.
• Cavernous hemangioma is a rare congenital lesion that appears commonly as a solitary vascular lesion of limited size.
• Racemouse hemangiomatosis may or may not be associated with systemic disease.
• A retinal vasoproliferative tumor is a solitary yellow-to-pink vascular mass usually in the inferior retina.