Choroidal and Retinal Tumors in Adults: 
Choroidal Metastases and Vascular Tumors of the Choroid

The conclusion of a four-part series on guidelines for diagnosing and treating ocular lesions.

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The first three articles of this four-part series discussed vascular tumors of the retina, choroidal melanocytic lesions, intraocular lymphoma, retinal astrocytoma, and choroidal osteoma. In this fourth and final article, we describe the clinical features, diagnosis, and treatment of choroidal metastases and of two types of vascular tumors of the choroid.

CHOROIDAL METASTASES

Metastatic cancer is the most common secondary intraocular malignancy. Most intraocular metastases are carcinomas, and they principally affect the choroid. Breast cancer in women and lung cancer in men are probably the most frequent primary tumors. Sarcomas and melanomas are less common. Most patients present with a single tumor in only one eye; however, careful examination often shows other probable small lesions. Local treatment can be effective, but the overall prognosis is usually poor.

Clinical features. Choroidal metastases tend to affect the posterior pole. They are often associated with exudative retinal detachment leading to metamorphopsia and vision loss, which are the most common ocular symptoms. Infiltration of the optic nerve is less common. Lesion color can vary, depending on the origin of the primary tumor: yellow (lymphoma and breast, lung, and gastrointestinal tract tumors), reddish-orange (renal cell carcinoma), reddish-orange to pink (follicular carcinoma of the thyroid), and pink or golden-orange (carcinoid).1,2

Diagnostic approaches. Fluorescein angiography (FA) usually shows late pinpoint hyperfluorescence. Indocyanine green (ICG) angiography often reveals subtle, occult lesions separate from the symptomatic lesion.3 Ultrasound usually shows high internal reflectivity and no prominent vascular pulsations. Orbital computed tomography (CT) and magnetic resonance imaging (MRI) can confirm the presence of a mass, but these diagnostic tests are not as useful for differential diagnosis. Full-body CT and MRI scans can help determine a primary mass if none is known. If a primary mass cannot be found, fine needle aspiration biopsy can be used in some cases with no history of previous extraocular cancer.4

Differential diagnosis. Differential diagnosis includes amelanotic nevus and melanomas, hemangioma, choroidal osteoma, primary choroidal lymphoma, astrocytoma, sclerochoroidal calcification, central serous chorioretinopathy, uveal effusion syndrome, vitelliform dystrophy, posterior scleritis, Harada disease, and tuberculosis.

Management. The principal options for managing intraocular metastases include radiotherapy, chemotherapy, and hormonal therapy.2 Small, asymptomatic tumors should be followed periodically.

CHOROIDAL HEMANGIOMA

Choroidal hemangiomas are rare, benign vascular tumors that occur in diffuse and circumscribed forms. On histology, these tumors appear as collections of thin-walled and variably sized vessels that may be predominantly cavernous, predominantly capillary, or of a mixed type. Diffuse hemangiomas are often associated with systemic disorders whereas circumscribed hemangiomas are not.5

Diagnosis. Fundus examination demonstrates a unilateral orange-to-red choroidal mass with poorly defined margins. Exudation can be present. A circumscribed hemangioma can occur over a background of a diffuse choroidal hemangioma. ICG angiography shows the typical intrinsic vascular pattern of this lesion. There is an initial hyperfluorescence followed by a washout effect. Ultrasonography examination can be useful. A dome-shaped choroidal mass with indistinct margins is observ-
able on B-scan, and high internal reflectivity is visible on A-scan. Macular edema can be detected and measured with optical coherence tomography (OCT).

**Differential diagnosis.** It is important to differentiate circumscribed choroidal hemangioma from malignant choroidal melanomas and choroidal metastases.

**Treatment.** Treatment options include observation, laser photocoagulation, radiotherapy, transpupillary thermotherapy, and photodynamic therapy. Photodynamic therapy has become the treatment of choice for this disease.

**DIFFUSE CHOROIDAL HEMANGIOMA**

The diffuse choroidal hemangioma is normally unilateral and ipsilateral to a facial hemangioma (nevus flammeus); these tumors are commonly seen in patients with Sturge-Weber syndrome.

**Diagnosis.** Fundus examination reveals a diffuse orange choroidal mass. A washout effect is not present on ICG angiography. Persistence of hyperfluorescence throughout the phases of the angiogram is common. A-scan detects high internal reflectivity. An associated circumscribed hemangioma can be present as well.

**Treatment.** These lesions are a challenge to treat; therefore, follow-up should be performed by a combination of glaucoma and retinal or oncology experts.

**CONCLUSION**

As stated at the outset of this four-part series, the period of life when cataracts develop coincides with the age when a higher prevalence of ocular tumors emerge. It is vital that cataract surgeons be able to recognize ocular tumors in their patients, distinguish these from other lesions, and refer for ocular oncologic care when appropriate.

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