Although vitreoretinal lymphoma, retinal astrocytoma, and choroidal osteoma are rare diseases, they do occur. Therefore, it is important to have clinical suspicion of these entities because proper diagnosis may have important systemic and ocular implications. The general ophthalmologist must stay forever vigilant for causes of visual loss that might not be attributable to anterior segment findings.

In the first two articles of our four-part series, we discussed vascular tumors of the retina and choroidal melanocytic lesions. In this third article, we describe the clinical features, diagnosis, and treatment of intraocular lymphoma, retinal astrocytoma, and choroidal osteoma.

INTRAOCULAR LYMPHOMA

Intraocular lymphoma is the infiltration of the uvea, retina, vitreous, or optic disc by malignant lymphoid cells. Vitreoretinal lymphomas, which account for most intraocular lymphomas, are high-grade, B-cell malignancies that are usually associated with central nervous system (CNS) disease, presenting before or simultaneously with primary vitreoretinal lymphoma (PVRL; previously called primary intraocular lymphoma), or after CNS involvement.\(^1\) Primary choroidal lymphomas are less common and tend to be low-grade with better systemic prognosis.

Clinical features. PVRL are usually bilateral and often present with vitritis and subretinal perivascular infiltrates, which frequently evolve to retinal pigment epithelium (RPE) crumpling, on ophthalmologic examination (Figure 1).\(^2\) Patients with choroidal lymphoma initially present with multifocal areas of whitish yellow coloration and spotting.

Diagnostic approaches. Diagnostic approaches for patients with intraocular lymphoma include vitrectomy, vitreoretinal biopsy, fine-needle aspiration biopsy, and transscleral choroidal biopsy.\(^3\)

Differential diagnosis. The differential diagnosis includes choroidal hemangioma, choroidal osteoma, amelanotic choroidal melanoma, metastatic carcinoma to the choroid, pneumocystis carinii choroiditis, other microbial choroiditis, posterior scleritis, Harada disease,
uveal effusion syndrome, and bilateral diffuse uveal melanocytic proliferation. Patients with PVRL usually have a marked disparity between vision and the amount of cells in the vitreous cavity. With so many cells in the vitreous, an eye with uveitis would have markedly worse vision than is seen with PVRL.

**Management.** The principal management options for intraocular lymphoma include external beam radiation therapy and chemotherapy (mainly methotrexate alone or in combination with rituximab).4 PVRLs are high-grade, large B-cell malignancies. Despite improvements in diagnosis and treatment, PVRL remains an aggressive disease with median progression-free survival of 29.6 months and overall survival of 58 months, unaffected by treatment type.4,5 Primary choroidal lymphoma is a low-grade lymphoma, and it may have a more favorable prognosis than PVRL. In these cases, radiotherapy has shown excellent results.

**RETINAL ASTROCYTOMA**

Retinal astrocytomas are rare, benign tumors that can be locally aggressive if they enlarge and begin to exudate. These lesions have a juxtapapillary location in many patients. In most cases, they are associated with tuberous sclerosis.

**Diagnosis.** An intraretinal whitish-yellow tumor is found on ophthalmoscopy. In some cases, well-defined nodules are observed (Figure 2). Fluorescein angiography (FA) demonstrates a superficial and deep intraretinal vascular network. Ultrasonography is a useful diagnostic test. A-scan shows medium to high internal reflectivity, and B-scan shows a homogeneous and nodular mass. In some cases, focal calcifications are detected.

**Differential diagnosis.** It is important to differentiate retinal astrocytoma from retinoblastoma. Differential diagnoses include retinal hemangioblastoma, granuloma, and amelanotic choroidal melanoma.

**Treatment.** In most cases, only observation is required. Shields et al5 reported resolution of exudation in retinal astrocytomas with the use of photodynamic therapy.

**CHOROIDAL OSTEOMA**

Choroidal osteoma is an ossifying benign tumor that tends to affect young women. This tumor is typically unilateral with a juxtapapillary location. Fundus examination demonstrates a low-lying, whitish-pinkish, well-delineated, plateau-shaped lesion. Large vessels can be seen within the lesion similar to those seen in Haversian canals. Indocyanine green (ICG) can help delineate these vessels. In some cases, choroidal neovascularization (CNV) may appear, and this can be detected on FA and ICG.7 Ultrasonography and computed tomography scans may show a calcified plaque at the choroid (Figure 3).

**Differential diagnosis.** The main differential diagnosis is sclerochoroidal calcification, which tends to occur in older persons, has a mulberry-type appearance, and is usually present outside the arcades.

**Treatment.** In most cases, only observation is required. If CNV appears, treatment should be performed. Photodynamic therapy and antiangiogenic agents, or a combination of the two, are generally good treatments. Sometimes photodynamic therapy can cause decalcification of the mass, which may result in visual loss.

**CONCLUSION**

Intraocular lymphoma, retinal astrocytoma, and choroidal osteoma are rare lesions. However, it is important to be aware of their clinical features to be able to...
perform an adequate diagnosis. In fact, intraocular lymphoma is an aggressive malignancy that can dramatically affect the patient’s life.

The final installment in our four-part series will cover choroidal hemangioma and choroidal metastases.

Luis Amselem, MD, is a Consultant in the Department of Ophthalmology at the Hospital Moises Broggi, Barcelona, Spain. Dr. Amselem states that he has no financial interest in the material presented in this article. He may be reached at e-mail: luisamselem@yahoo.es.

Teresa Diago, MD, is a Consultant in the Department of Ophthalmology at the Hospital de Sagunto, Valencia, Spain. Dr. Diago states that she has no financial interest in the material presented in this article. She may be reached at e-mail: tediasem@gmail.com.

Jose S. Pulido, MD, MPH, MS, MBA, is a Professor in the Department of Ophthalmology at the Mayo Clinic, Rochester, Minnesota. Dr. Pulido states that he has no financial interest in the material presented in this article. He may be reached at e-mail: pulido.jose@mayo.edu.