

The Seasick Pattern of Choroidal Lymphoma on Enhanced Depth Imaging Optical Coherence Tomography

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Lymphoma can involve several sites in the periocular region, including the conjunctiva, eyelid, orbit, and tissues within the eye. Intraocular lymphoma is generally classified into either vitreoretinal or uveal lymphoma. Vitreoretinal lymphoma is aggressive, with vitreous, retinal, optic nerve, and brain involvement, and often requiring systemic and intrathecal chemotherapy. Uveal lymphoma is more indolent and can involve the iris, ciliary body, and, most commonly, the choroid, often with related conjunctival or orbital involvement.¹ Choroidal lymphoma typically manifests as a unilateral condition with multifocal sites of infiltration scattered throughout the fundus and with minimal subretinal fluid.¹

Choroidal lymphoma, most often classified as extranodal marginal zone lymphoma, is considered to be a low-grade malignancy with a favorable prognosis.^{1,2} In some cases, this condition can be subtle and the tumor imperceptible, leading to misdiagnosis as uveitis or central serous chorioretinopathy.³ In such cases, imaging with enhanced depth imaging optical coherence tomography (EDI-OCT) can be helpful to detect subclinical infiltration.³ Here we describe a case of subtle choroidal lymphoma with identifying features on EDI-OCT imaging.

CASE REPORT

A 68-year-old white man noticed painless subtle blurred vision in his left eye (OS) over 2 years. He has previously undergone retinal detachment repair in the

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right eye (OD) with resultant 20/80 visual acuity. On examination 18 months prior to referral, a thin choroidal lesion 7 mm in diameter was noted. At 10 months prior to referral, a second lesion 4 mm in diameter was observed. Systemic evaluation was unremarkable.

Upon referral, visual acuity was 20/80 OD and 20/25 OS. Intraocular pressures were 18 mm Hg OD and 16 mm Hg OS. The anterior segment was unremarkable in each eye. On ophthalmoscopy, peripheral chorioretinal scarring from the previous retinal detachment was found in the right eye, and there was no choroidal infiltration. The left fundus revealed a diffuse, ill-defined, orange-yellow equatorial choroidal mass 14 mm in diameter. Overlying retinal pigment epithelium (RPE) alterations and shallow subretinal fluid were present. On autofluorescence photography, hyperautofluorescence was noted, suggestive of lipofuscin clumping (Figure 1). Ocular ultrasonography disclosed

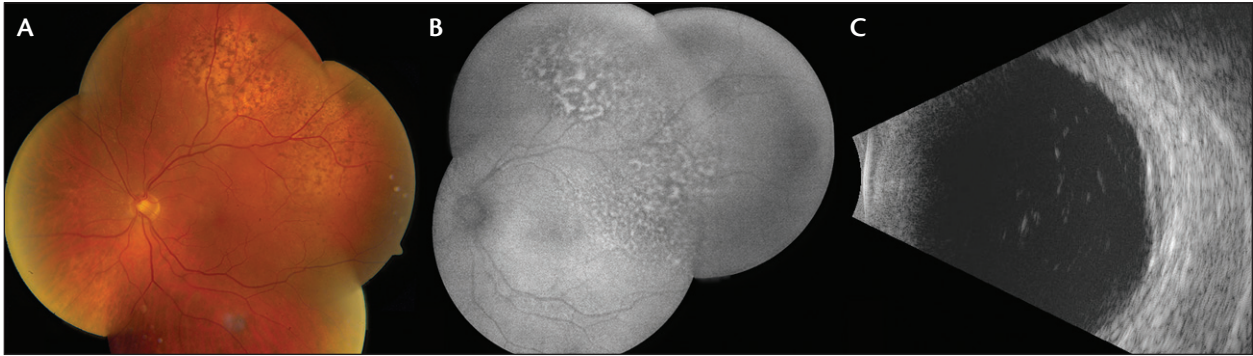


Figure 1. An ill-defined choroidal lymphoma in a 68-year-old man (A). A diffuse choroidal mass superotemporally with overlying lipofuscin pigment clumping was confirmed on autofluorescence (B). Ultrasonography depicts the minimally thickened choroidal mass without extrascleral extension (C).

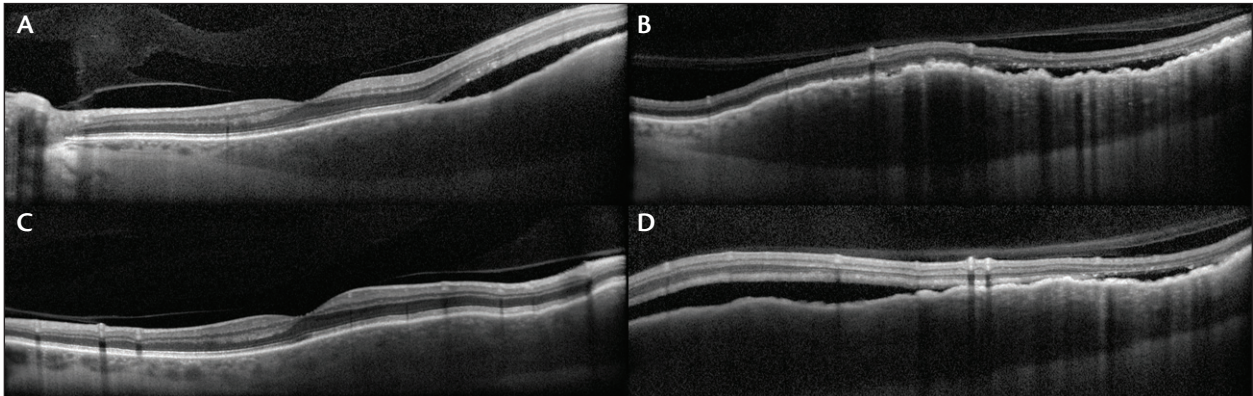


Figure 2. EDI-OCT scans of choroidal lymphoma. A foveal image depicts intact retina and slightly undulating choroidal surface contour with deep choroidal infiltration (A). Overlying the main portion of the tumor, there is subretinal fluid with photoreceptor loss, and the choroid is markedly thickened with deep shadowing (B). Note the irregular clumping of presumed macrophages on the surface of the RPE. In other areas, the tumor shows a seasick undulating surface (C). Overlying subretinal fluid is seen, and the photoreceptor band is broadened (D).

a diffusely thickened choroid with an acoustically hollow mass 3 mm in thickness showing medium to low internal reflectivity.

On EDI-OCT, there was an irregular, undulating appearance to the choroidal surface that has been described as seasick,³ with compression of the choriocapillaris inward (Figure 2). Other features on EDI-OCT included deep optical shadowing and numerous clumps of optically dense material at the level of the RPE, presumed to represent macrophages with lipofuscin. Overlying subretinal fluid with a homogeneous, optically dense, and widened photoreceptor band in some areas was noted, as was photoreceptor loss in other areas.

These features were suggestive of choroidal lymphoma. Fine-needle aspiration biopsy was considered to confirm the diagnosis but was not performed because this tumor involved the patient's better-seeing eye. The patient was followed conservatively. Continued

enlargement of the mass with an increase in subretinal fluid necessitated treatment using oral prednisone (60 mg daily), which was tapered slowly over 5 months. External radiotherapy was avoided because, again, this was the patient's better-seeing eye. At 16 months follow-up, the subretinal fluid was resolved, and visual acuity remained 20/25 OS with a stable intraocular tumor.

DISCUSSION

Choroidal lymphoma can present with a discrete amelanotic intraocular mass, or it can manifest as a subtle, ill-defined choroidal thickening without obvious mass. This more subtle presentation can be challenging to recognize and can lead to erroneous diagnoses of presumed central serous chorioretinopathy, atypical uveitis, and others.²⁻⁴ In an evaluation of 73 eyes with choroidal lymphoid neoplasia in 59 patients, the referral diagnosis was correct in 27 (46%) patients.⁴ The erroneous referral

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diagnoses included choroidal metastasis (5%), melanoma (3%), unknown choroidal tumor (20%), and others (7%). In that cohort, patients' symptoms included blurred vision (64%), pain (5%), periocular redness (7%), foreign body sensation (3%), and proptosis (2%). In 17% of cases there were no symptoms.

Choroidal lymphoma generally appears as an orange-yellow single or multifocal mass elevating the RPE. The infiltration can be a solitary focal mass (15%), multifocal patchy infiltration (25%), diffuse confluent infiltration (27%), or mixed (27%).⁴ Other features include choroidal folds (38%), subretinal fluid (48%), and optic disc edema (6%). A classic feature of uveal lymphoma is transscleral epibulbar tumor extension (34%), appearing as a salmon-colored mass deep to Tenon fascia. Ultrasonography is particularly useful for identifying and measuring the extent of uveal infiltration and for detecting the extrascleral component.⁴

EDI-OCT provides valuable high-resolution cross-sectional details of the retina, choroid, and, in some cases, sclera.⁵ This capability allows characterization of intraocular tumors, particularly those of retinal and choroidal origin, and it is particularly helpful for cases in which the tumor is clinically undetectable.⁶ Features of choroidal nevus, melanoma, metastasis, osteoma, and other conditions based on EDI-OCT have been described.^{6,7} Regarding choroidal lymphoma, Shields et al found that EDI-OCT reveals infiltration in the outer choroid with inward compression of the choroidal tissue, imparting a variety of tumor surface topographies: a placid appearance with thin infiltration of 1 to 2 mm; a rippled appearance with infiltration of 2- to 3-mm thickness; and a seasick or undulating appearance with infiltration of 3 to 4 mm or greater.⁸

In the present case, the lymphoid infiltration was clinically evident, and EDI-OCT disclosed an undulating (rippled) contour in some areas and seasick contour in thicker areas. The choroidal vascular details were masked by the infiltration. Overlying subretinal fluid was evident, and hyperreflective micronodularity on the surface of the RPE probably represented macrophages with lipofuscin, as seen on autofluorescence.

EDI-OCT shows different surface contours for various intraocular tumors. Choroidal melanoma and nevus appear as discrete, smooth, dome-shaped surface topography, whereas metastasis is more irregular,

described as *lumpy bumpy*.^{7,9,10} Choroidal lymphoma shows variable configuration depending on the thickness, appearing flat if infiltration is thin, rippled when thicker, and seasick when even thicker.⁸ In our case, EDI-OCT provided important information regarding the extent and surface topography of infiltration. This modality is also useful in patient follow-up after therapy for documenting subretinal fluid resolution and tumor response. ■

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