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Looking Beyond and Behind a Retinal Detachment

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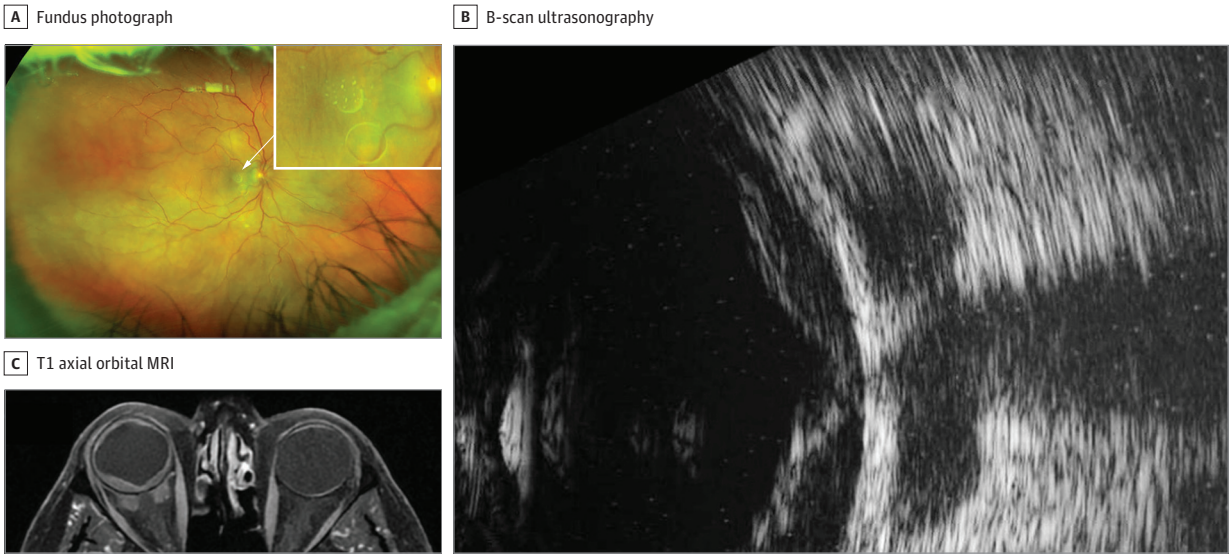

Figure 1. A, Fundus photograph of the right eye showing an infratemporal exudative retinal detachment. Inset, Choroidal folds in the macula. B, B-scan ultrasonography demonstrating broad areas of low reflectivity within the sub-Tenon space and around the optic nerve in addition to the presence of a retinal detachment. C, T1 axial orbital magnetic resonance image (MRI) with gadolinium showing the right eye retinal detachment and revealing an enhancing right optic nerve mass.

A 63-year-old woman with a history of type 1 diabetes presented with gradually progressive vision loss in the right eye for the past year. Her visual acuity on presentation was 20/80 OD and 20/25 OS. Anterior examination findings of both eyes were normal with clear posterior chamber intraocular lenses. Fundus examination of the right eye showed choroidal folds and a macula-involving retinal detachment with no identifiable retinal break and shifting subretinal fluid (**Figure 1**A). Fundus examination findings in the left eye were normal. Fluorescein angiography did not identify areas of leakage. B-scan ultrasonography demonstrated broad areas of low reflectivity within the sub-Tenon space and around the optic nerve (Figure 1B). Magnetic resonance imaging (MRI) revealed a contrast-enhancing area of soft-tissue thickening encircling the right optic nerve (Figure 1C). Serologies for syphilis, Lyme disease, rheumatoid factor, and antineutrophilic cytoplasmic antibody–associated vasculitis were negative. Chest radiographic findings were unremarkable.

WHAT WOULD YOU DO NEXT?

A. Begin oral prednisone

B. Observe with interval orbital MRI

C. Perform pars plana vitrectomy for retinal detachment repair

D. Perform biopsy of the optic nerve lesion

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Diagnosis

Low-grade B-cell lymphoma associated with exudative retinal detachment

What to Do Next

D. Perform biopsy of the optic nerve lesion

Discussion

This case represents orbital lymphoma masquerading as posterior scleritis, both of which may cause choroidal folds, exudative retinal detachments, and sub-Tenon fluid. Given the lack of retinal break, vitrectomy for repair of the retinal detachment would not have addressed the underlying etiology (answer C). B-scan ultrasonography in this patient's case revealed a focus of low reflectivity adjacent to the optic nerve (Figure 1B), which was felt to be atypical of posterior scleritis and prompted orbital MRI that showed soft-tissue thick-

ening around the optic nerve (Figure 1C). After negative results on infectious and inflammatory workup, differential diagnosis included idiopathic orbital inflammation (IOI), lymphoma, or other lymphoproliferative processes. Whereas IOI typically presents acutely and with pain, the course of this patient's vision loss was insidious and nonpainful. Moreover, IOI is a diagnosis of exclusion. Empirical treatment for IOI with oral prednisone could reduce biopsy yield in the case of lymphoma or lymphoproliferative processes (answer A). Given the potentially life-threatening nature of orbital lymphoma, observation with interval orbital MRI would not be appropriate (answer B).

Positron emission tomography did not reveal extraorbital lesions amenable to biopsy. Consequently, orbital biopsy was performed via transconjunctival incision with takedown of the medial rectus muscle (answer D). Prior to surgery, extensive discussion with the patient covered procedural risks, including severe vision loss. The orbital biopsy specimen underwent histologic correlation and flow cytometry, which identified low-grade B-cell lymphoma.

Although rare, orbital lymphomas are the most common type of orbital malignant neoplasm, usually presenting as a low-grade B-cell lymphoma or extranodal marginal zone lymphoma of the mucosa-associated lymphoid tissue type.[1] Classically, it presents as an insidiously progressive and painless orbital process in middle-aged and older individuals.[2] While early diagnosis of orbital lymphoma is critical for the preservation of vision, delay of diagnosis can occur due to nonspecific orbital symptoms among orbital inflammatory and neoplastic processes.[3] Despite advances in using conventional MRI features and imaging-related values to differentiate orbital lymphoma from orbital inflammatory processes,[4] currently, adequate imaging studies followed by early surgical biopsy are key for achieving early diagnosis.[5]

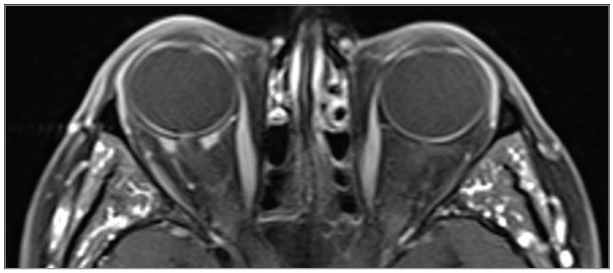

Figure 2. T1 axial orbital magnetic resonance image with gadolinium following radiotherapy treatment of the right orbit, demonstrating an interval size reduction of the enhancing optic nerve mass and resolution of the retinal detachment.

On diagnostic confirmation by the pathologist, patients should be sent to the oncologist and/or radiation oncologist for consultation. Excellent local control may be achieved for low-grade lymphomas with a conventional radiotherapy dose ranging from 24 to 40 Gy. Alternatively, recent studies suggest that a low-dose course (defined as 2 Gy × 2 fractions) with salvage therapy in the case of locoregional relapse may achieve comparable outcomes, although further multicenter studies are warranted.[6,7]

Patient Outcome

This patient underwent low-dose radiotherapy treatment to the right orbit, which led to reduction of the optic nerve mass and resolution of the retinal detachment (**Figure 2**). This was associated with improvement in visual acuity to 20/25 OD. This case demonstrates the vital role imaging plays in ruling out tumors as a cause for exudative retinal detachment masquerading as posterior scleritis.

ARTICLE INFORMATION

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