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Orbital Lymphoma Masquerading as Panuveitis

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Abstract

Purpose—To describe a case of orbital mucosa-associated lymphoid tissue (MALT) lymphoma masquerading as unilateral panuveitis.

Methods—Retrospective chart review.

Results—A 53-year-old female patient with unilateral vitritis and exudative retinal detachment refractory to immunosuppressive treatment was eventually diagnosed with orbital MALT lymphoma. Following treatment with radiotherapy and rituximab, the patient's intraocular inflammation and retinal detachment resolved.

Conclusions—Orbital MALT lymphoma can masquerade as refractory unilateral panuveitis with exudative retinal detachment and appears to respond to a combination of radiotherapy and specific B-cell-targeted systemic therapy.

Keywords

MALT lymphoma; orbital MALT lymphoma; Rituximab; uveitis; masquerade syndrome

Masquerade syndromes are noninflammatory disorders that initially present as ocular inflammation.¹ Identification of these patients is imperative due to the high frequency of underlying malignancy that can impact not only visual outcome but also systemic morbidity and mortality.

Lymphomas can masquerade as any type of ocular inflammation with an initial favorable response to corticosteroids but frequently the “inflammation” recurs. The majority of patients with orbital lymphoma present with a palpable mass or proptosis.^{2,3} This report describes the clinical presentation and course of a patient with orbital mucosa-associated lymphoid tissue (MALT) lymphoma presenting as panuveitis.

CASE REPORT

A 53-year-old white female with a 1-year history of uveitis and progressive vision loss in the right eye was referred to the National Eye Institute with a visual acuity of 20/80 OD and 20/16 OS. She was HLA-B27 positive and had a history of sacroiliitis. Ophthalmic exam showed 2+ cells and flare in anterior chamber, trace vitreous cells and haze, an inferior serous retinal detachment (RD), diffuse depigmentation resembling a sunset glow fundus,

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macular edema, and papillitis OD (Figure 1A). Fluorescein angiography (FA) showed disc leakage OD, B-scan ultrasound demonstrated diffuse choroidal thickening with Tenon's cysts. Ocular coherence tomography confirmed macular edema and subretinal fluid OD (Figure 1B). Examination and ancillary tests were normal OS. Initial laboratory investigations were unremarkable (complete blood count (CBC), chem20, erythrocyte sedimentation rate (ESR), creative protein (CRP), angiotensin converting enzyme (ACE), anti-neutrophil cytoplasmic antibody (ANCA), Lyme, chest radiography, purified protein derivative (PPD), rapid plasma reagin (RPR), syphilis IgG, serum/urine electrophoresis). Limited aqueous humor specimen measured below detectable levels of IL-6 and IL-10.

The patient was treated with prednisone (0.5 mg/kg/day) for presumed noninfectious panuveitis with resolution of the anterior chamber (AC) inflammation and unchanged vitritis. Mycophenolate mofetil and cyclosporine were subsequently added for 6 months without any significant change. A workup for masquerade syndrome, including computerized tomography of the chest, abdomen, and pelvis, mammography, and whole-body gallium-67 scintigraphy, was unremarkable. Cerebral spinal fluid was negative for infectious etiologies, flow cytometry, and cytology. Immunofixation electrophoresis demonstrated an IgG peak. MRI showed orbital soft tissue thickening with enhancement of the posterior sclera and intraorbital optic nerve (Figure 2A). Immunosuppressive therapy was subsequently tapered, orbitotomy with orbital and optic nerve sheath biopsy was consistent with MALT lymphoma (Figure 2B). Polymerase chain reaction (PCR) was positive for IgH gene rearrangement and negative for *Helicobacter pylori* DNA. Following treatment with orbital radiotherapy (total 3960 cGy) and rituximab infusions, the intraocular inflammation and the serous RD resolved with residual atrophic retina, a best corrected visual acuity (BCVA) of 20/50 OD, and a myopic shift consistent with resolution of the serous RD (Figure 1C, D).

DISCUSSION

This case demonstrates an orbital MALT lymphoma masquerading as refractory unilateral panuveitis treated with radiation and rituximab. MALT lymphoma is a B-cell lymphoma seen in older individuals and has an indolent course and systemic association in 20% of cases.³ Gaucher et al. described 3 cases of ocular MALT lymphoma masquerading as posterior uveitis or scleritis resistant to prednisone.⁴ The gastric variety of MALT lymphoma has been associated with *H. pylori*.^{1,2} However, no globally accepted prognostic or infectious factors exist for ocular MALT lymphoma.⁵ In contrast to nodal lymphomas, low-grade MALT lymphomas respond favorably to local treatments, but radiotherapy alone risks relapse.^{1,2} Chemotherapy is often reserved for patients with systemic disease.²

In this case, optic nerve swelling, choroidal thickening, and serous RD were likely secondary to a direct infiltrative process of the optic nerve and choroid with secondary uveal effusion. Although the initial clinical presentation was panuveitis, the response to therapy was unsatisfactory. Masquerade syndrome should be suspected in any patient who presents with uveitis that does not show the expected response to therapy.

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FIGURE 1. Fundus photographs and OCT images before and after treatment for MALT lymphoma. Initial examination showed a depigmented fundus with optic nerve head swelling (A) and macular edema on OCT (B). Following treatment, as the serous RD resolved the fundus appeared more depigmented, accompanied with pigmentary deposits in the macula, but the optic nerve swelling and macular edema improved (C, D).

FIGURE 2. MRI brain/orbits and histopathology of orbital MALT lymphoma. T1-weighted axial images with fat suppression and gadolinium demonstrated abnormal enhancement within the orbit, along the posterior sclera and intraorbital optic nerve (A). Photomicrograph of orbital MALT lymphoma demonstrates many small round and cleaved atypical lymphoid cells surrounding the nerve (B).