Case Report

Idiopathic Orbital Inflammation Associated with Panuveitis in an Adult and a Review of the Literature

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Abstract

Purpose: To report an unusual case of idiopathic orbital inflammation associated with panuveitis.

Case report: We report a case of a 47-year-old female with a history of uveitis who presented with abrupt and painful proptosis with complete vision loss in short period and was unresponsive to antibiotic therapy. B-scale ultrasonography and magnetic resonance both showed retinal detachment and orbital space-occupying mass. The patient underwent orbitotomy and the diagnosis of idiopathic orbital inflammation was confirmed by histopathology. Typically, anterior uveitis is rarely seen in idiopathic orbital inflammation in adults. Here, we report an unusual case of idiopathic orbital inflammation associated with panuveitis, which has not been reported in literature so far.

Conclusion: Inflammation involving both the eye and the orbit is rarely seen in adults. Idiopathic orbital inflammation and panuveitis might share a similar mechanism in this case. (***Eye Science 2013; 28:–***)

Keywords: orbital inflammatory disease; idiopathic orbital inflammation; panuveitis; adult; surgery

Introduction

Idiopathic orbital inflammation (IOI), also known as orbital pseudotumor, is the third most common cause of orbital inflammation, ranking after thyroid-associated orbitopathy and lymphoproliferative disease1. The peak incidence is in the 4th to 5th decades of life[1]. Since this process can involve any orbital structure or cell type, it has widely varying clinical presentations, with the most common ones of proptosis, pain, swelling, palpable mass, and motility restriction[2,3]. The pathogenesis of IOI remains elusive, but autoimmunity has been suggested as a possible reason and associations have been reported between IOI and some systemic disorders such as Wegener's granulomatosis, eosinophilia, giant-cell arteritis, polyarteritis nodosa, Crohn's disease, and sclerosis[4,5]. To our knowledge, orbital inflammation associated with Behçet's disease without ocular involvement (retinal vasculitis or uveitis) has been reported in only three cases[6-8]. Anterior uveitis has been described previously as a rare manifestation of IOI in a few pediatric patients and in only one adult so far9. In this report, we describe an unusual case of an adult with idiopathic orbital inflammation associated with panuveitis.

Case Report

A 47-year-old female presented with abrupt and painful proptosis with complete vision loss in the left eye for 20 days. She also experienced redness, lid swelling, ptosis, and tearing, but had no fever or chills. Antibiotic therapy was ineffective. She reported having a history of uveitis of her left eye for one year. Topical and systemic corticosteroids and immunosuppressants were used irregularly with her vision had declined gradually to 20/200 before the onset of proptosis. She also had a history of tympanitis of both ears and severe intracranial infection secondary to ear surgery performed 10 years previously.

Physical examinations showed a normal state of consciousness and normal body temperature and pulse; her blood pressure was 151/95 mmHg. The patient had no light perception in the left eye The intraocular pressure and orbital pressure were both moderately raised on finger palpation. Redness and massive firm swelling of the lids were obvious, with high skin temperature and 3-mm dysraphism. Slit

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lamp examination revealed conjunctival redness, edema, prolapse, and a visible and palpable subconjunctival firm mass. Keratic precipitates were positive; corneal epithelial defects and exposure corneal ulcer were detected in the inferior third of the cornea. Anterior chamber flare and activity cells were both graded 2+ without hypopyon. Posterior synechiae and pupillary seclusion prevented posterior segment examination. Orbital pressure graded 3+ and eyeball fixation was noted. Exophthalmometer examination showed a 7 mm greater projection of the affected eye than the fellow eye (Figure 1). The visual acuity in the fellow eye was 1.0. Slit lamp and fundus examinations were unremarkable.

White blood cell and neutrophil counts were 12.3×10^9/L and 8.9×10^9/L, respectively. B-scale ultrasonography showed retina detachment with a characteristic "T" sign, and an orbital space-occupying mass with effusion. Magnetic resonance (MR) showed an orbital space-occupying, soft tissue mass with a molding growth pattern, as well as retina detachment (Figure 2). Since corticosteroids and immunosuppressants, as well as antibiotic therapy, were of limited effectiveness in this patient, immediate orbitotomy was performed to exclude malignant orbital diseases. We used an anterior route with a superior and transconjunctival approach to remove the visible mass that filled the entire retrobulbar and periocular space. The diagnosis of idiopathic orbital inflamma-

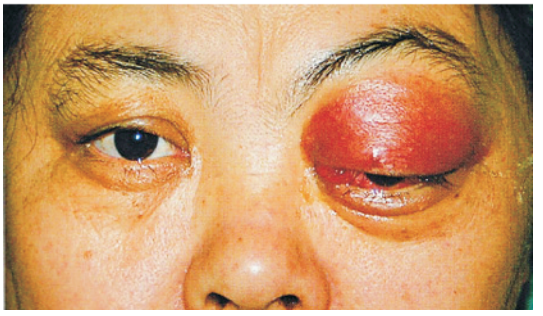

Figure 1 Preoperative photograph shows proptosis, ptosis, lid swelling, and redness in the left eye.

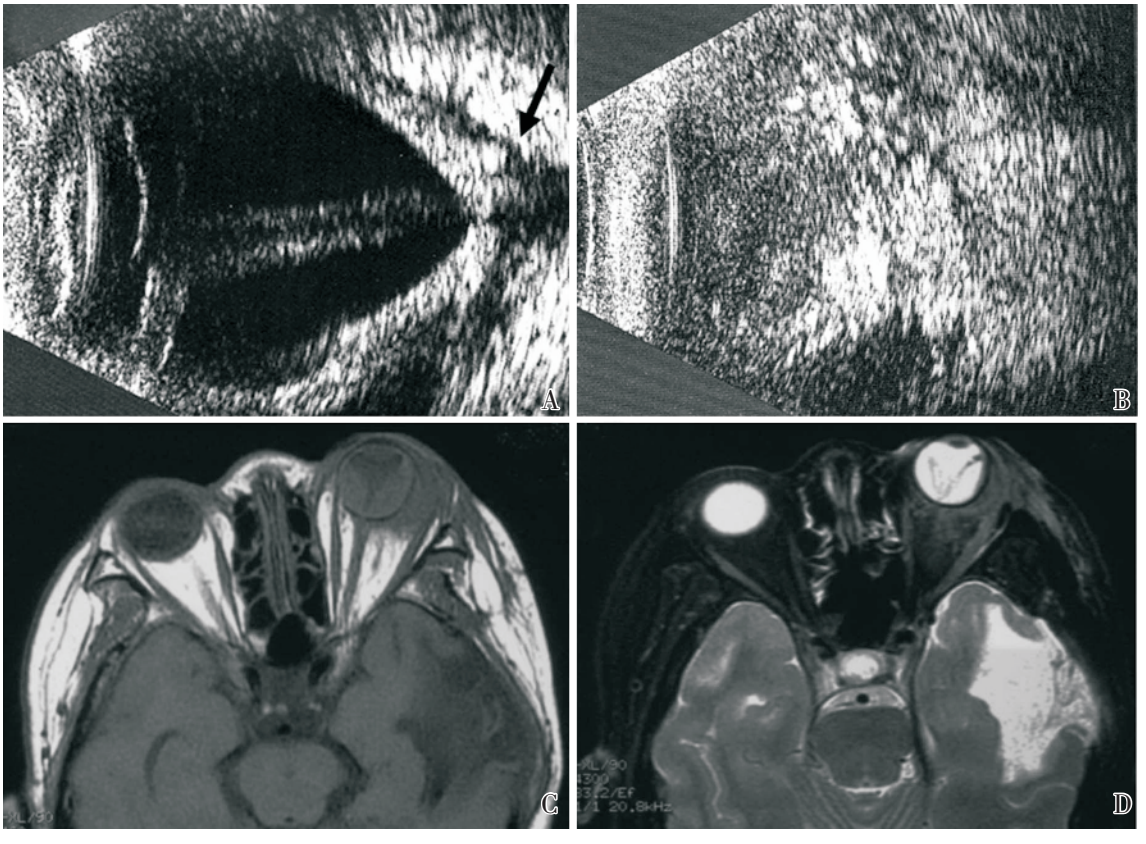

Figure 2 Image manifestations of the left eye. A, B-scale ultrasonography shows retina detachment with a characteristic "T" sign (arrow). B, B-scale ultrasonography shows orbital space-occupying mass with effusion. C, Axial T1-weighted MRI shows an orbital space-occupying mass-like soft tissue with molding growth pattern, retina detachment, and an old cerebromalacia. D, Image of axial T2-weighted MRI.

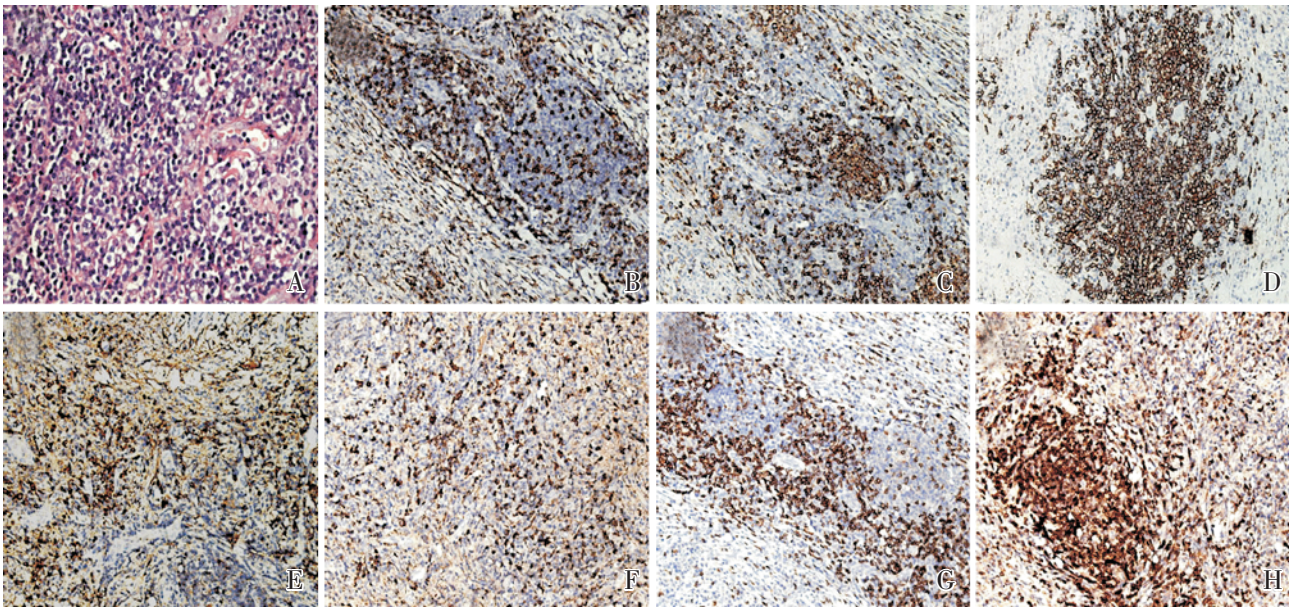

Figure 3 Idiopathic orbital inflammation, lymphocyte hyperplasia subtype, A, Hematoxylin and eosin stain, magnification X 40, shows proliferation of lymphocytes and plasmocytes. B, Positive CD45Ro staining. C, Positive CD79 staining. D, Positive CD20 staining,. E, Positive Kappa staining. F, Positive Lambda staining. G, Positive CD3 staining. H, Positive Bcl–2 staining. B~H, magnification ×20.

tion with lymphocyte hyperplasia subtype was confirmed by histopathology and immunohistochemistry (Figure 3).

Discussion

The associations of anterior uveitis and idiopathic orbital inflammation have been reported in several pediatric patients[10-14]. Additionally, Pang et al. reported a 35-year-old male with acute anterior uveitis, optic disc swelling, posterior scleritis, and orbital inflammatory disease, which responded well to systemic corticosteroids[9]. Idiopathic orbital inflammation associated with systemic Behçet's disease without ocular involvement has been described previ ously in only three patients. Two of these were Behçet's disease patients with myositis involving the orbit[6,7], and the other one manifested lacrimal gland involvement[8].

Several features of the clinical presentations were notable in our patient. 1) The process from the onset to complete vision loss was rather acute (less than 20 days), which was rarely seen in typically self-limited, isolated idiopathic orbital inflammation. The abrupt and active inflammatory phase of panuveitis may cause not only anterior segment inflammation but also total retina detachment and even quick proliferative vitreoretinopathy. The manifestation of ultrasonography and MRI in our case coincided with this pathological process. Once the detached retina "stuck" together, complete vision loss could occur. 2) The patient had a one-year history of uveitis. Despite her gradual vision loss, the process was relatively slow and controllable. The concurrence of vision loss and proptosis suggested that the inflammation process was activated by similar mechanism that caused both ocular and orbital involvement. 3) Clinical deterioration continued despite the use of systemic and topical steroids and immune suppressants. For financial reasons, the patient was not monitored at long-term follow-up after she was prescribed the above mentioned drugs. Relapse might be related to irregular or inadequate usage of the drugs. 4) The orbital mass involved retrobulbar space as well as subconjunctiva space, which was similar to the characteristic growth pattern of lymphoma or other lymphocyte proliferation diseases as a mass molding to the shape of the globe[9]. In this case, however, the onset was acute, which was not consistent with the typical clinical course of lymphoma. Even so, serious and life-threatening diseases (for example, lym-

phoma) should be excluded by immediate emergent biopsy or orbitotomy. The following treatment of this patient should involve systemic and topical anti-inflammatory agents. Once the inflammatory process is quiescent, vitrectomy and silicone oil filling might be considered to prevent atrophy of eyeball.

In conclusion, idiopathic orbital inflammation may uncommonly manifest itself together with panuveitis in adults. Idiopathic orbital inflammation associated with panuveitis should be one of the essential considerations in unusual cases with both ocular and orbital inflammation.

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