Contralateral Recurrence of Idiopathic Orbital Inflammatory Syndrome in a Pediatric Patient

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An 11-year-old African American male presented to our emergency department (ED) with complaints of progressive headache and vomiting. His symptoms included right eye swelling and pain with eye movement, conjunctival injection and decreased visual acuity. Review of systems was negative for diplopia, trauma, fevers, neck stiffness, weight loss, joint pain and rashes. Past medical history was notable for steroid-responsive orbital pseudotumor in the left eye, which was diagnosed by CT two years earlier (Figure 1). Physical examination revealed a vitally stable patient with right-sided periorbital edema, proptosis and ptosis with conjunctival injection over the lateral orbit. Extraocular movements showed limitation of right upgaze compared with the left and pain with eye movement in all directions (Figure 2). Pupils were equally reactive and fundus examination was normal. Visual acuity without the patient’s eye glasses was 10/25 in the right and 20/100 in the left.

Laboratory evaluation included normal electrolytes, renal function, transaminases and complete blood count. C-reactive protein was elevated (3.64 mg/dL) as was sedimentation rate (59 mm/hr). Total IgG was normal (1,480 mg/dL), and IgG subclasses revealed an elevated IgG1 (981 mg/dL, normal 456-952) and IgG4 (175 mg/dL, normal 1-168). Angiotensin-converting enzyme was normal (32 U/L). FANA screen, perinuclear and cytoplasmic ANCA, Quantiferon Gold and lyme serology were negative. MRI was consistent with idiopathic orbital inflammation (IOI), demonstrating edema of the right periorbital tissues, retroconal fat and extraocular muscles (Figure 3).

The patient was started on methylprednisolone with dramatic clinical improvement (Figure 4). He was discharged on a steroid taper with prostaglandin eye drops. On follow-up with rheumatology, he was noted to have recurrent pain and was started on mycophenolate mofetil with subsequent resolution of symptoms.

Discussion

Idiopathic orbital inflammation, also known as orbital pseudotumor, is a benign condition characterized by orbital inflammation without evidence of systemic disorders. Although an underlying autoimmune cause is suspected, the etiology remains elusive. It is characterized by ophthalmoplegia, conjunctival injection, pain, proptosis, and/or ptosis. Although IOI is the third most common cause of orbital inflammation in adults following Grave’s disease and lymphoproliferative disorders, pediatric cases are much less common. Systemic corticosteroids are first-line treatment, and an abrupt clinical response is thought to be pathognomonic. Disease recurrence occurs in up to 37% of adults. However, the rate of recurrence among children is unknown.

This patient was diagnosed with IOI and subsequent recurrence in the contralateral orbit two years after initial diagnosis. To our knowledge, there are only three reports in the literature describing IOI recurrence among children. Yan et al. described one case of recurrence in a series involving 24 children. That patient was treated with corticosteroids but required radiotherapy to achieve complete recovery. Belanger et al. described 12 children with IOI, three of whom had recurrence after corticosteroid therapy. Belanger et al. described 12 children with IOI, three of whom had recurrence after corticosteroid therapy. The site of recurrence was not described in either report. Finally, Avni-Zauberman et al. described two pediatric cases (a 17- and an 18-year-old) of IOI with recurrence in the contralateral orbit, termed “migratory” disease. To our knowledge, our patient is the youngest described case of disease recurrence in the contralateral orbit. Interestingly, our patient had an elevated IgG4,
which has been described in two pediatric case reports.\(^8,9\) The role of IgG4 in disease pathogenesis is currently unclear. MM

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REFERENCES


Learning points

- IOI is a rare disease in children.
- Diagnostic work-up includes MRI or CT and laboratory studies to rule out thyroid disorders, oncologic disease (rhabdomyosarcoma) or autoimmune diseases (sarcoidosis, Wegener's).
- Treatment includes systemic corticosteroids but may involve radiotherapy or immunosuppressive agents in refractory cases. Biopsy is rarely needed and typically based on treatment response.

FIGURE 1

Computed tomography showing proptosis and postseptal inflammatory changes involving the retroconal fat and lateral rectus muscle of the left orbit.

FIGURE 2

Magnetic resonance image of the right orbit demonstrating proptosis and marked edema of the retroconal fat and lateral rectus muscle.

FIGURE 3

Initial exam showing periorbital edema of the right eye, with proptosis, ptosis and conjunctival injection over the lateral aspect of the orbit. There was limitation of upgaze on the right.

FIGURE 4

Subsequent examination (12 hours later) after a single dose of methylprednisolone showing dramatic improvement in periorbital edema, ptosis and improving upgaze.