Painless orbital myositis

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Abstract

Idiopathic orbital inflammation is the third most common orbital disease, following Graves orbitopathy and lymphoproliferative diseases. We present a 11 year old girl with 15 days history of painless diplopia. There was no history of fluctuation of symptoms, drooping of eye lids or diminished vision. She had near total restricted extra-ocular movements and mild proptosis of the right eye. There was no conjunctival injection, chemosis, or bulb pain. There was no eyelid retraction or lid lag. Rest of the neurological examination was unremarkable.Erythrocyte sedimentation rate was raised with eosinophilia. Antinuclear antibodies were positive. Liver, renal and thyroid functions were normal. Antithyroid, double stranded deoxyribonucleic acid and acetylcholine receptor antibodies were negative. Repetitive nerve stimulation was negative. Magnetic resonance imaging (MRI) of the orbit was typical of orbital myositis. The patient responded to oral steroids. Orbital myositis can present as painless diplopia. MRI of orbit is diagnostic in orbital myositis.

Key Words

Idiopathic orbital inflammation, orbital pseudotumor, orbital myositis, painless

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Introduction

Orbital myositis (OM) is a common variant of idiopathic orbital inflammatory disease (IOID). IOID is also known as orbital pseudotumor. IOID is the third most common orbital disease, following Graves orbitopathy and lymphoproliferative diseases.[1] OM implies orbital inflammation confined to one or more extraocular muscles. It usually begins acutely with unilateral periorbital pain, particularly with eye movement and diplopia. In addition, signs of ocular inflammation like eyelid swelling and conjunctival injection are usually present.[2] The diagnosis can be made clinically due to the characteristic pain and signs of inflammation. T2-weighted magnetic resonance imaging (MRI) of the orbit shows swelling and hyperintensity in the extraocular muscles.[3] We present an unusual case of OM with subacute-onset painless diplopia without signs of ocular inflammation. Investigations for thyroid orbitopathy (TO), systemic autoimmune diseases, orbital neoplasm and ocular myasthenia were negative. Typical features of OM were detected on MRI of the orbit.

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Case Report

A 11-year-old girl presented with complaints of binocular diplopia in all directions since 15 days. There was no history of headache, orbital pain or painful eye movements. There was no history of fluctuation of symptoms, drooping of eyelids or diminution of vision. There was no history of fever, rash, joint pain or any other systemic symptoms. There was no history of preceding infection or trauma to the eye. There was no history of weakness in limbs. There was no family history of similar complaints.

General examination was normal. On neurologic examination, cranial nerves were normal except extraocular movements. The extraocular movements in the right eye were restricted in all gazes, except possible mild elevation and depression. There was horizontal diplopia in all gazes. There was mild proptosis in the right eye. There was no conjunctival injection, chemosis or bulb pain. Visual acuity, visual fields, pupillary reaction, fundoscopy and eyelids were normal. There was no eyelid retraction or lid lag. There was no audible bruit on auscultation over the right eye.

The intraocular tension in both eyes was normal (14.6 mmHg). Rest of the neurological examination was unremarkable. A differential diagnosis of TO, lymphoproliferative infiltration, orbital neoplasm, OM and ocular myasthenia was considered.

The total leukocyte count was normal (9800/mm^3), with eosinophilia (14%). The erythrocyte sedimentation rate was

elevated (70 mm/1 h). Liver, renal function and thyroid functions were normal. Antithyroglobulin (ATA) and antithyroid peroxidase antibodies (ATPA) were negative. Antinuclear antibody was positive (1:100), while antibodies to double-stranded deoxyribonucleic acid were negative. Acetyl choline receptor (AChR) antibodies and the repetitive nerve conduction studies were negative.

MRI of the orbit showed swelling and uniform hyperintensity on T2 images in the right superior, inferior, medial and lateral recti muscle involving the belly and tendon insertion [Figure 1a] with minimal postcontrast enhancement suggestive of OMShe was started on oral prednisolone 20 mg/day, which resulted in prompt resolution of symptoms and hyperintense signal [Figure 1b]. Steroids were tapered over the next 4 weeks. There is no recurrence of symptoms at 6 months of follow-up.

Discussion

OM constitutes 29% cases of IOID.[4] OM is clinically characterized by acute-onset orbital pain, impairment of ocular movement and diplopia. Proptosis, swollen eyelids, scleritis and conjunctival hyperemia may be seen depending on the extent of the inflammation. Pain, typically worsening with eye movement, is observed in 85–100% of the cases.[2,5] Our patient did not have ocular pain and presented as subacute onset of painless diplopia. The extraocular muscle paresis did not follow a pattern of III, IV, VI nerve palsy and, hence, an extraocular muscle, neuromuscular junction disease was suspected. TO, lymphoproliferative disorders, orbital neoplasm, myasthenia gravis and cavernous sinus disease (CSD) were thought as possibilities in addition to a restricted form of OM.

TO is the most common orbital disease,[6] and is the most important differential diagnosis of OM.[7] It has a similar presentation with diplopia, restricted ocular motility and proptosis. TO is usually insidious, with bilateral (85%) or unilateral (10–30%) proptosis, conjunctival injection, periorbital edema, eyelid retraction and lid lag.[6] Muscle insertions are typically spared in TO as compared with OM, where they are involved. A normal thyroid profile and negative antithyroid antibodies (ATA, ATPA) ruled out TO in our case. Among the orbital neoplasms, rhabdomyosarcoma is the most common childhood orbital neoplasm. It usually presents as rapidly progressive proptosis with displacement of eye, diplopia and restricted ocular motility. The course was not progressive in our patient and proptosis was mild, unlike rhabdomyosarcoma. The absence of signs of inflammation and periorbital pain made us think about ocular myasthenia, although there was no fluctuation of symptoms, fatigability and strictly unilateral involvement. AChR antibodies, repetitive nerve stimulation and edrophonium tests were negative. CSD can mimic OM.[8] CSD affects oculomotor nerves, unlike in our patient, where individual extraocular muscles were involved. Carotidcavernous fistula was unlikely as there was no episcleral, conjunctival venous congestion nor pulsating exophthalmos with elevated intraocular tension.

There was eosinophilia with an absolute count of 1372 cells/µl. Although eosinophilia is commonly associated with Churg-Strauss syndrome[8] and eosinophilic myositis, there were no features suggestive of these diseases in our patient. She did not have any features of systemic autoimmune diseases like systemic lupus erythematosus, rheumatoid arthritis or sarcoidosis. Presence of eosinophilia can be a clue of OM in children, and is seen in 31.03% of the cases.[9]

MRI showed hyperintense signal in the right superior, inferior, medial and lateral recti muscles [Figure 1a] with minimal enhancement, suggestive of myositis. The superior and inferior oblique muscles were spared. Oblique muscles are the least commonly affected muscles in OM, as seen in our patient. The absence of pain may be attributed to a less-severe form of the disease termed limited oligosymptomatic ocular myositis.[10] Here, the inflammation is mild and restricted to few ocular muscles. Patchy involvement of extraocular muscles and lack of involvement of other orbital structures can lead to a painless presentation and lack of signs of ocular inflammation.

Conclusion

Orbital myositis can present as painless diplopia without signs of ocular inflammation. MRI of orbit is diagnostic of orbital myositis.

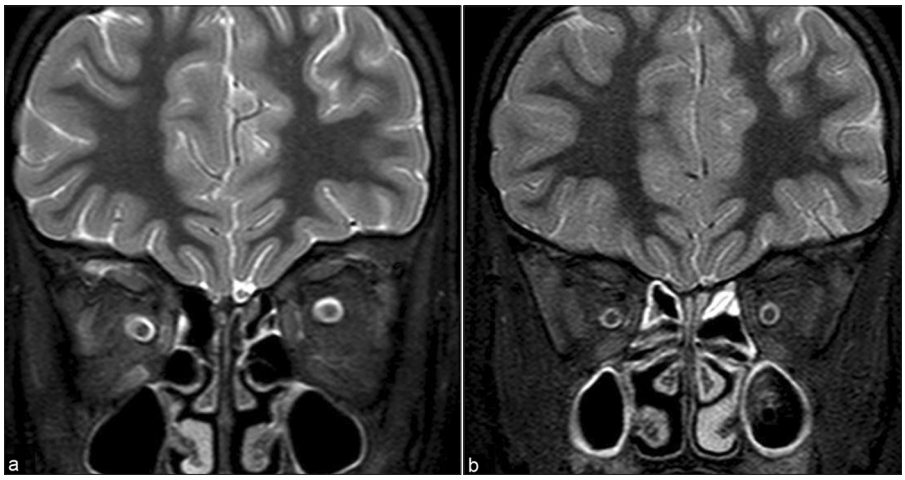

Figure 1: (a) T2-weighted coronal image showing hyperintense signal in the right inferior, superior, lateral and medial recti muscles, suggestive of orbital myositis, (b) Resolution after treatment with oral corticosteroids

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