• Letter to the Editor •

Ocular myositis—a diagnostic dilemma

Ishita Bajaj, Jolly Rohatgi

University College of Medical Sciences, New Delhi 110095, India

Correspondence to: Ishita Bajaj. B-1/462, Janakpuri, New Delhi 110058, India. i.bajaj90@gmail.com

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Dear Editor,

Orbital myositis is a relatively rare entity but is the next most common cause of extra ocular muscle disease after thyroid associated orbitopathy (TAO)[1-2]. It was first described by Gleason in 1903 as an orbital pseudotumour and then renamed in 1930[3]. It usually affects young females than males. Extraocular muscles differ from skeletal muscles by a smaller unit size and higher motor neuron discharge, higher blood flow and volume of mitochondria fractions that allow inflammatory cells to reach and circulate easier, causing inflammation[4].

Clinical findings include moderate to severe orbital pain, painful diplopia increased with eye movement, exophthalmos, swollen eyelids, conjunctival hyperemia or chemosis usually. Contrast enhanced magnetic resonance imaging (CEMRI) is the most sensitive modality which discloses enhancement or thickening of inflamed extraocular muscle.

The study was conducted in accordance with the principles of the Declaration of Helsinki. The informed consent was obtained from the subject.

A 35 years old male patient presented to the outpatient department with a complaint of gradually increasing proptosis in the right eye over the past 1mo. There was a history of tooth extraction followed by pain around 2wk prior to this episode. There was no associated history of trauma or weight loss. There was no history of any systemic disease.

On examination, the patient was afebrile with no evidence of infections, lymphadenopathy or masses.

Assessment of the right eye showed that there was an axial proptosis of 4 mm. Visual acuity was 6/6, with an intraocular pressure of 29 mm Hg. There was mild limitation in superior gaze and moderate in lateral gaze in the involved eye, with a central Hirschberg test (HBT; Figure 1).

There was also a 2 mm upper lid ptosis present. The episcleral vessels were also injected and tortuous in the right eye especially temporally and superiorly. Pupil was 5 mm, sluggishly reacting on right side although consensual reflex was present.

Fundus examination of the affected eye revealed dilated and tortuous vessels with disc hyperemia, rest of the fundus was normal.

Left eye examination was normal with 6/6 vision, and intraocular pressure of 17 mm Hg.

All blood investigations including thyroid function test, inflammatory markers and blood sugars were within normal limits.

A contrast enhanced computed tomography (CECT) scan was done which showed involvement of both belly and tendinous insertions of superior and medial rectus muscles but there was no evidence of cavernous sinus thrombosis. CEMRI showed thickening of medial and superior rectus on the right side with post contrast enhancement likely to be inflammatory associated with cavernous sinus and perisellar region enhancement (Figure 2).

On this basis the patient was started on intravenous antibiotics, low dose heparin and antiglaucoma drugs. Further an orbital Doppler was done which showed prominent long ciliary arteries. Central retinal, ophthalmic, long ciliary arteries showed normal waveform and no occlusion was seen.

On initial examination because of the gaze restrictions and ptosis, a diagnosis of partial 3rd plus 6th nerve palsy with cavernous sinus thrombosis was made. But radiological investigations ruled out cavernous sinus thrombosis as the cause of nerve palsies. Also, the patient did not respond to antibiotics. Thyroid ophthalmopathy was also ruled out because of the euthyroid status. Therefore, a differential of orbital myositis was made although the picture was not very clear. Initially the patient was started on intravenous antibiotics, low dose heparin and antiglaucoma drugs.

After 2wk of antibiotics, pupil reaction became normal but there was only a mild decrease in congestion. No other improvement occurred and therefore oral steroids were also started.

Extraocular movements and proptosis started improving slowly after 1wk of starting steroids. Pupil was normal reacting. IOP started lowering. Although the episcleral venous congestion and mild ptosis still persisted.

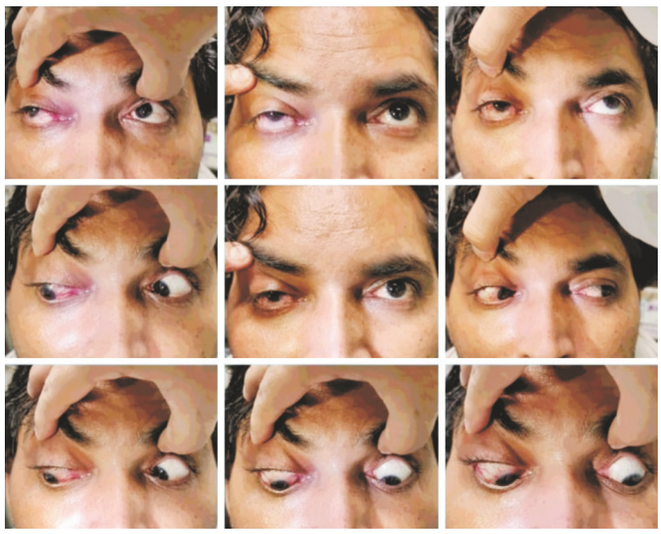

Figure 1 Proptosis and a lateral and superior gaze limitation in the right eye on presentation.

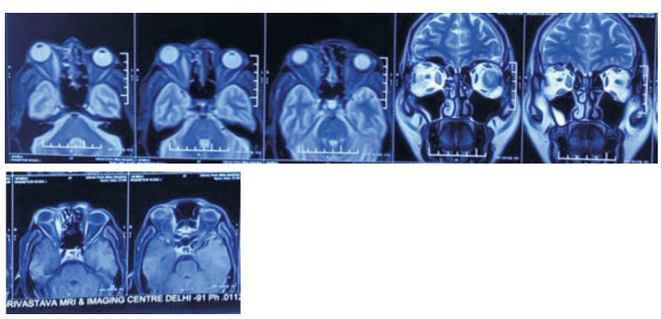

Figure 2 Axial and coronal MRI sections show bulky medial and superior rectus muscles with enhancement of cavernous sinus.

After a month of giving oral steroids, proptosis decreased to 2 mm. The extraocular movements were now full and free. Pupil was normal reacting. Although mild episcleral venous congestion and mild upper lid ptosis was still present. Steroids were tapered very slowly over a period of 6wk. At present, the patient is not on any medications and the eye examination is now within normal limits.

DISCUSSION

Today, orbital myositis represents a subgroup within the wide entity of idiopathic orbital inflammatory syndrome also known as orbital pseudotumour[5]. It's a rare inflammatory disorder which can involve a single or multiple extraocular muscles, usually affects young females more than males (2:1). Initial presentation include acute to subacute painful diplopia most commonly. There are two major forms 1) a limited oligosymptomatic ocular myositis (LOOM) with additional conjunctival injection only, 2) a severe extensive exophthalmic ocular myositis (SEOM) with additional ptosis, chemosis and proptosis[5].

It is defined as an idiopathic inflammation of the extraocular muscles in the absence of thyroid disease, ocular myasthenia gravis, and other systemic, particularly autoimmune mediated diseases, resembling $CD4^+$ T cell-mediated dermatomyositis[5]. It is usually a diagnosis of exclusion after patient's laboratory tests have been obtained and physical examination has been done.

Contrast enhanced orbital magnetic resonance imaging is the most sensitive modality disclosing thickening and post contrast enhancement of the affected muscle including the myotendinous insertion as well as shown in this case[6].

Typically, oral corticosteroids in the dose of 1–1.5 mg/kg•d for 1–2wk remain the mainstay of treatment with a slow taper over 6–12wk. In severe cases pulse intravenous methylprednisolone can be administered for 3d before shifting to the above mentioned regime[7-8]. Whereas in atypical, recurrent cases or cases with severe side effects to steroids, steroid sparing immunosuppressants can be given. Antimetabolites, such as azathioprine, methotrexate, mycophenolate mofetil are frequently used immunosuppressives in this condition.

Extraocular myositis is a rare inflammatory disorder which needs to be diagnosed and treated promptly with oral steroids which is usually the first line of treatment unless contraindicated or in case severe side effects. It is usually a diagnosis made after ruling out all other causes with similar clinical findings with the help of history, laboratory tests and radiological imaging where contrast enhanced orbital MRI is the most sensitive modality available.

ACKNOWLEDGEMENTS

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