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The diagnostic puzzle of orbital myositis

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Abstract

Background: Idiopathic orbital inflammatory disease (IOID) is a diagnosis of exclusion that poses a diagnostic challenge as it may closely resemble other orbital pathologies.
Case report: A 69-year-old woman presented with progressive blurred vision in the left eye associated with eye pain, redness, proptosis and restricted extraocular movements for 3 weeks. Orbital imaging was suggestive of thyroid orbitopathy, but thyroid function tests were normal. After 8 weeks of medical decompression and oral prednisolone with a suboptimal response, an incisional biopsy of the left lateral rectus muscle revealed non-malignant skeletal muscle with inflammatory cell infiltration, consistent with IOID. Systemic corticosteroid therapy was continued for a total of 16 weeks, resulting in significant improvement in visual acuity, proptosis, and extraocular muscle function.
Conclusion: Given that IOID can mimic orbital pathologies, appropriate imaging and histopathological confirmation are essential for accurate diagnosis. Timely and adequate corticosteroid therapy can lead to favourable visual and functional outcomes.

Keywords: idiopathic orbital inflammatory disease, muscle biopsy, orbital myositis, orbital pseudotumor

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Cabaran diagnostik myositis orbital

Abstrak

Pengenalan: Penyakit radang orbital idiopatik (IOID) merupakan diagnosis pengecualian yang menimbulkan cabaran diagnostik kerana ia hampir menyerupai patologi orbital lain.
Laporan kes: Seorang wanita berusia 69 tahun hadir dengan penglihatan kabur progresif pada mata kiri, dikaitkan dengan sakit mata, kemerahan, bengkak dan pergerakan mata terhad selama 3 minggu. Imbasan orbital menunjukkan orbitopati tiroid, namun ujian fungsi adalah normal. Selepas 8 minggu rawatan dekompresi dan prednisolon oral dengan respons yang kurang memuaskan, biopsi pada otot 'lateral rectus' kiri menunjukkan tisu otot rangka tidak malignan dengan infiltrasi sel radang, selaras dengan IOID. Terapi kortikosteroid sistemik diteruskan selama 16 minggu, menghasilkan peningkatan ketara dalam ketajaman penglihatan, bengkak, dan fungsi otot ekstraokular.
Kesimpulan: Memandangkan IOID boleh menyerupai patologi orbital lain, pengimejan yang sesuai serta pengesahan histopatologi adalah penting untuk diagnosis yang tepat. Rawatan kortikosteroid yang tepat pada masanya dan mencukupi boleh membawa kepada hasil visual dan fungsi yang baik.

Kata kunci: biopsi otot, miositis orbital, penyakit radang orbital idiopatik, pseudotumor orbital

Introduction

Idiopathic orbital inflammatory disease (IOID), also known as pseudotumor, is a benign inflammatory disorder of unknown origin within the orbit.[1,2] IOID represents approximately 8–10% of orbital mass lesions and is the third most common orbital disease after thyroid orbitopathy and lymphoproliferative diseases.[1] It is commonly observed in middle-aged individuals, with no sex predilection.[1] IOID is classified based on the site of involvement, encompassing anterior, diffuse, apical, posterior, myositis, dacryoadenitis, periscleritis, perineuritis, and focal mass subtypes.[1]

Clinically, IOID presents with various ocular symptoms, with eye pain and periorbital swelling being common.[1] While most cases of orbital myositis can be diagnosed clinically without the need for a biopsy, certain atypical cases such as this case require a biopsy for accurate diagnosis. This article presents a case of orbital myositis mimicking thyroid ophthalmopathy and neoplasm, highlighting diagnostic challenges and the critical role of biopsy.

Case presentation

A 69-year-old woman with underlying hypertension and bilateral primary open-angle glaucoma presented with progressively worsening blurred vision in the left eye (OS) associated with binocular diplopia, eye redness, periorbital swelling, and restricted eye movement for 3 weeks. She denied photophobia, lacrimation, or headache. She also denied ocular trauma, connective tissue disease, thyrotoxicosis, fever, infective symptoms, or constitutional symptoms.

OS visual acuity (VA) was 6/60, (pinhole: 6/24) with a positive relative afferent pupillary defect. The OS was proptosed, hypotropic, and with periorbital swelling (Fig. 1, top). The conjunctiva was injected, chemosed inferiorly, the cornea was clear, and the anterior chamber was deep and quiet. Extraocular movement (EOM) was significantly limited in all gazes. Fundus showed tortuous vessels; otherwise, the optic disc was pink, not swollen, with a cup-to-disc ratio of 0.5, retina was flat, no choroidal folds, and no macular abnormalities. Intraocular pressure was normal. Hertel exophthalmometry measurement showed OS at 22 cm and OD at 20 cm. OD VA was 6/9 with no abnormalities seen. Systemic examination was unremarkable, including no palpable thyroid.

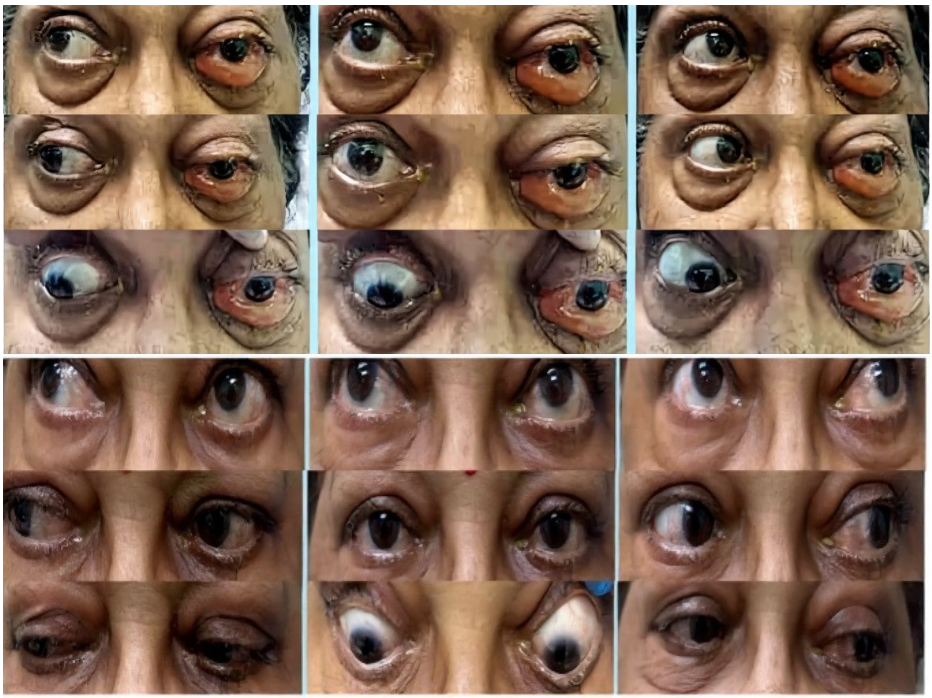

Fig. 1. (Top) Left eye proptosis with periorbital swelling and restricted EOM. *(Bottom)* Left eye post-treatment resolution of proptosis and improved EOM.

Blood investigations, including a full blood count, coagulation profile, inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein, antinuclear antibody, rheumatoid factor, tumour markers, thyroid hormone, thyroid peroxidase antibody, and syphilis and viral screenings were normal. The magnetic resonance imaging (MRI) scan showed a central, haemorrhaging, fusiform enlargement of the OS lateral rectus muscle measuring 3 cm (anteroposterior) x 2 cm (width) x 2 cm (craniocaudal), sparing the tendon and displacing the OS optic nerve (Fig. 2).

Intravenous 1 g methylprednisolone was administered daily for 3 days, followed by oral prednisolone with an initial dose of 1 mg/kg/day (a total of 60 mg/day), then tapered to 10 mg weekly for a total of 40 mg, and by 5 mg weekly for a total of 20 mg over the course of 8 weeks. A repeat scan showed no reduction in the lesion's size. The patient was co-managed with the oculoplastic team, and a left lateral rectus incisional biopsy demonstrated non-malignant skeletal muscle with inflammatory cells, discounting a diagnosis of neoplasm (Fig. 3). Oral prednisolone was furthered tapered to 2.5 mg weekly for a total of 5 mg for another 8 weeks. While on steroid therapy, patient was also on calcium supplements and gastric protection, with stable blood glucose levels and no obvious weight gain noted. The patient exhibited excellent recovery with a significant improvement in unaided VA to 6/9, optic nerve function, and EOM (Fig. 1b). At the latest monthly follow-up, the patient had been in remission for 10 months.

Discussion

Given that the MRI scan showed an EOM belly enlarged without tendon involvement, potential differential diagnosis for this case was thyroid orbitopathy. A similar finding was seen in a study where 6 patients with orbital myositis showed no tendon involvement.[3] However, our patient exhibited no other signs of thyroid eye disease and was euthyroid, making the diagnosis of thyroid orbitopathy unlikely. The absence of preceding infection and leucocytosis ruled out orbital cellulitis.[4] The biopsy clearly ruled out a tumour, which was initially suspected due to the presence of haemorrhage in the muscle seen on the MRI scan. The haemorrhage in our case could have been associated with inflammatory changes and mechanical stress induced by the enlarged muscle.[5] Haemorrhage can also occur in cases of orbital varices, which present with similar symptoms.[6]

Early intervention improves outcomes.[7] An early incisional biopsy was performed to guide the management as the patient showed a poor response to initial steroid therapy. Biopsy is generally recommended in cases with progressive neurologic deficits, lack of steroid responsiveness, persistent imaging abnormalities, and recurrence.[1]

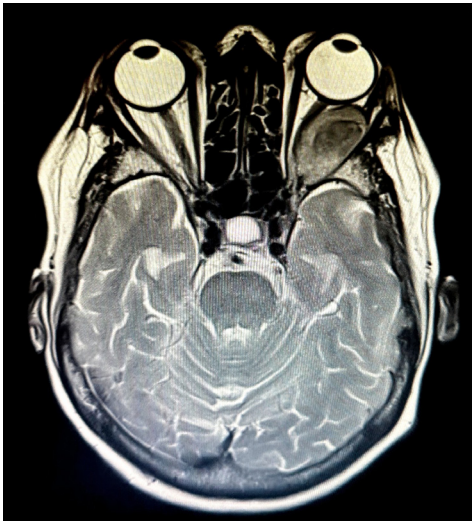

Fig. 2. MRI scan showing an enlarged left lateral rectus muscle.

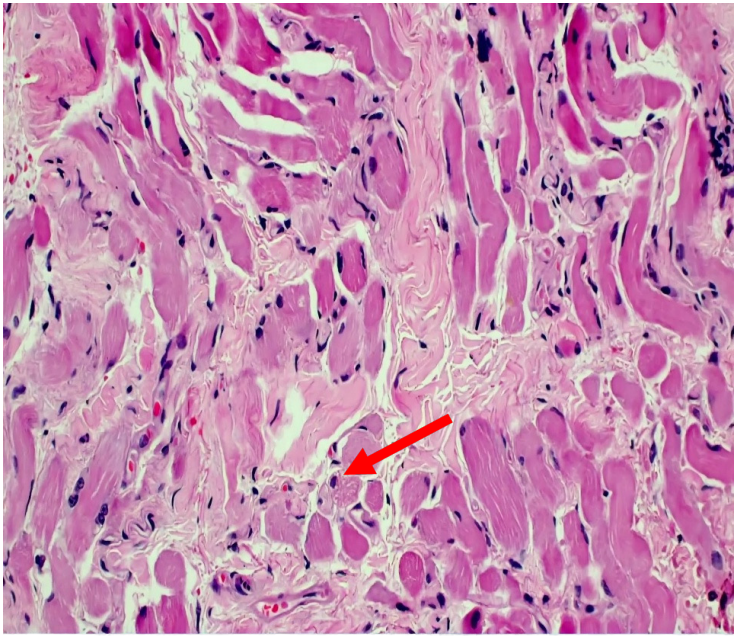

Fig. 3. Biopsy revealing benign skeletal muscle with inflammatory cells.

The primary aim of treatment is to reduce inflammation and improve vision. Corticosteroid therapy is the first-line treatment for orbital myositis. The typical initial dose is 1 mg/kg/day for a week, followed by a gradual tapering over 6–12 weeks.[8] In cases of severe presentation, intravenous methylprednisolone is prescribed for 3 days, followed by the previously described oral steroid regimen.[8] Since immunosuppressive therapy takes 6–8 weeks to manifest its full effect, maintenance of steroid therapy is necessary.[8] The primary mechanism of corticosteroids involves the inhibition of rapidly dividing cells, particularly leukocytes, thereby inducing an anti-inflammatory effect mediated by the inhibition of phospholipase A2 and cyclo-oxygenase pathways.[1]

Conclusion

This case highlights that prompt diagnosis, investigation, and management are crucial in achieving favourable outcomes in IOID.

Declarations

Informed consent for publication
The patient provided written informed consent for the publication of the clinical data and images provided in this article.

Competing interests
None to declare.

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