

IMAGES IN CLINICAL PRACTICE

MORE THAN MEETS THE EYE - A CASE REPORT OF ORBITAL PSEUDOTUMOR

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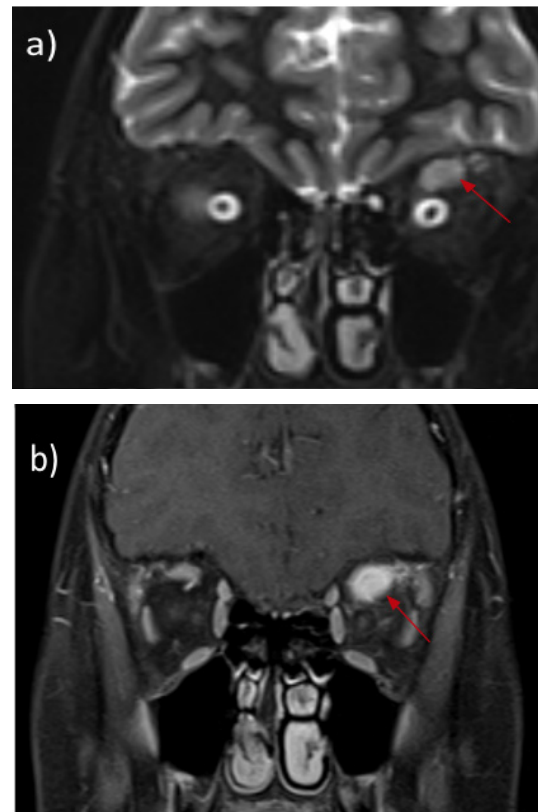
A previously healthy 10-year-old girl, presented with a 1-week history of holocranial headache, mostly affecting the frontotemporal area, coupled with photophobia, phonophobia and two episodes of vomiting. She also reported pain while looking downward and double vision when looking upward to the left. There was no fever, motor or sensory impairments or seizures. Upon admission, she exhibited left eye ptosis and convergent strabismus in the left eye. She also had difficulty in the upward left eye movement and experienced pain during downward left eye movement. She mentioned double vision when looking straight, upward and to the left, which resolved upon closing each eye individually. Pupils were isochoric and equally responsive to light. There were no other neurological signs. Furthermore, she didn't have conjunctival injection or chemosis, and the funduscopy examination was normal.

Blood tests had no signs of acute inflammation, elevated sedimentation rate, hyperuricemia, elevated lactate dehydrogenase or peripheral blast cells. Thyroid function was normal. Sarcoidosis was excluded (normal angiotensin-converting enzyme and chest radiograph). Immunological studies showed negative results for rheumatoid factor, antinuclear antibodies, anti-DNA antibodies and HLA-B27. Infectious study showed positive IgG for CMV, EBV, VZV and *Mycoplasma pneumonia*, but without positive IgM. *Borrelia burgdorferi* IgM and IgG were both negative. HIV was negative. Magnetic resonance imaging (MRI) displayed thickening of the superior rectus muscle, hyperintensity on T2-weighted images, and enhancement with gadolinium injection, without areas of diffusion restriction. Cranial MRI was normal.

What is the diagnosis?

This is a case of a myositis pseudotumor of the superior rectus muscle, that was causing restriction in ocular supraversion and adduction of the left eye because of inflammation on the superior rectus muscles that was contiguous with the lateral rectus muscle. She was treated with oral prednisolone (1 mg/kg), with improvement within 48 hours. The prednisolone was gradually tapered over 8

Figure 1. a) MRI T2 Short tau inversion recovery (STIR), coronal view; b) MRI T1 fat-suppressed (FS), after intravenous injection of gadolinium, coronal view. MRI shows thickening of the superior rectus muscle, hyperintensity on T2-weighted images and enhancement with gadolinium injection (red arrows).



weeks, with complete resolution of symptoms. Orbital pseudotumor, also known as Idiopathic Orbital Inflammatory Syndrome (IOI), is a benign, non-infectious, inflammatory orbital condition that poses challenges in both diagnosis and treatment. It contributes to 8 to 10% of all orbital masses.¹ It stands as the third most prevalent eye injury subsequent to thyroid orbitopathy and orbital lymphoma. Although infrequent among children, it can manifest at any age, gender or ethnicity.² According to its precise location, IOI can be categorized as anterior, diffuse, apical, posterior, myositis, or dacryoadenitis. Myositis may encompass one or

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multiple extraocular muscles. The medial rectus muscle is the most frequently affected. Clinically, it manifests with unilateral and periorbital pain, motility restriction, diplopia, ptosis, eyelid swelling, and conjunctival injection at the tendon insertion site.^{2,3} Constitutional signs and symptoms, including headache, fever, vomiting, anorexia, lethargy and abdominal pain, may be present in up to 50% of pediatric patients.⁴

The etiopathogenesis is currently unknown, but some authors suggest an immune-mediated mechanism following an acute infection. About 10% of patients present with an autoimmune disease^{5,6,7,8}, but it has also been associated with Streptococcal infection⁹, Lyme disease¹⁰ and Varicella-zoster disease.¹¹

The differential diagnosis encompasses acute orbital cellulitis, thyroid orbitopathy, orbital vasculitis, orbital trauma with retained foreign body, rhabdomyosarcoma, lymphoma, ruptured dermoid cyst, lymphangioma, Langerhans cell histiocytosis, secondary orbital retinoblastoma, Wegener granulomatosis and sarcoidosis.^{2,12} It is often misidentified as orbital cellulitis or an orbital mass with conjunctivitis. Given its potential impact on vision, prompt diagnosis and treatment are essential.

Laboratory tests are often normal, but may reveal peripheral eosinophilia, elevated sedimentation rate and elevated serum antinuclear antibodies. Imaging techniques help in the exclusion of alternative etiologies.² Myositis is distinguished from thyroid orbitopathy, which affects EOM bilaterally and spares the myotendinous junction with an increase in orbital fat volume.³ Myositis can be initially treated with systemic corticosteroids (prednisolone at 1-2 mg/kg per day), which typically results in symptom resolution within 24 to 48 hours, and is effective as a sole treatment in at least 40% of patients. When used with other immunosuppressants its efficacy is elevated to 60%. There is a potential role for intraorbital corticosteroids, either as a standalone treatment or in combination with systemic corticosteroids.³ In cases of treatment failure, contraindication to corticosteroids or recurrence, it is recommended to use low-dose radiotherapy² and consider orbital biopsy.¹³ Immunomodulatory agents and surgical resection ought to be reserved for refractory cases.³

Compliance with ethical standards

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