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Case Report

Idiopathic lateral rectus myositis in preteens: A case report [☆]

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ABSTRACT

Idiopathic orbital myositis is a subgroup of idiopathic orbital inflammation of unknown etiology that primarily involves extraocular muscles. Orbital myositis most commonly affects young adults in the third to fourth decade of life with a female predilection. Imaging is an important diagnostic technique. Inflammatory changes in the tendon and its insertion point to idiopathic orbital myositis. Other inflammatory, endocrine, or neoplastic diseases must be ruled out. We report a case of a 12-year-old girl who presented to the ophthalmologic emergency department with spontaneously developing pain during horizontal eye movement in the left eye and nonspecific symptoms, such as light sensitivity and slightly blurred vision. Her symptoms were nonspecific and physical examination was not diagnostic. However, computed tomography and magnetic resonance imaging of the head revealed isolated, monolateral, and monomuscular lateral rectus myositis as a manifestation of idiopathic orbital inflammation. The patient rapidly responded to systemic corticosteroids. Diagnostic radiology plays a significant role in the unsuspected diagnosis of myositis. Radiologists must be aware of this rare presentation of idiopathic orbital inflammation in preteens.

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Introduction

Idiopathic orbital inflammation (IOI) includes all nonspecific orbital inflammations of unknown etiology that can affect various orbital structures [1]. One of the subtypes of IOI is orbital myositis, also known as myositis pseudotumor that primarily affects extraocular muscles. It occurs most frequently

in young to middle-aged adults, with a 2-1 female predominance. However, the pathophysiology of this condition remains unknown. Patients typically present with orbital pain exacerbated by eye movements and diplopia. Other common findings include minimal proptosis, conjunctival injection, chemosis, and periorbital edema. Diagnostic imaging plays a significant role in the diagnosis. Corticosteroids are the most common first-choice therapy with good outcomes [2].

Abbreviations: IOI, idiopathic orbital inflammation; CT, computed tomography; MRI, magnetic resonance imaging; ENT, ear-nose-throat specialist.

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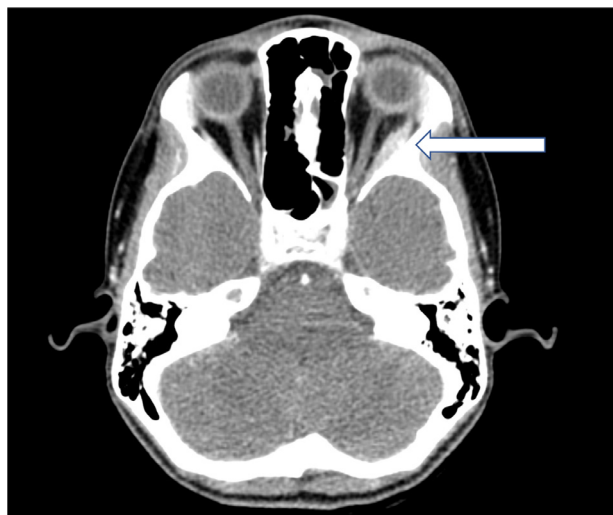


Fig. 1 – Contrast-enhanced CT scan, axial view, shows enlarged left lateral rectus muscle with contrast enhancement (arrow).

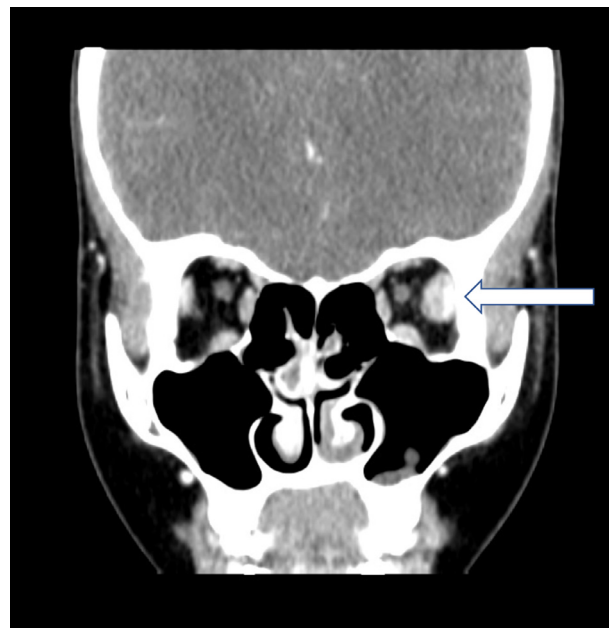


Fig. 2 – Contrast-enhanced CT scan, coronal view, shows enlarged left lateral rectus muscle with contrast enhancement (arrow).

Clinical case and imaging findings

A 12-year-old girl visited the ophthalmologic emergency department with spontaneously developing pain in the left eye for a day. The pain occurred during horizontal eye movement, especially on the right gaze associated with light sensitivity and slightly blurred vision. There was no pain in the primary eye position, double vision, or color vision abnormality. She complained of fatigue but was vitally stable and afebrile, with no headaches, neurological complaints, or history of trauma.

She had been using nasal spray for the last 2 days due to nasal congestion and was treated with antibiotics for tonsillitis by her family doctor about a month ago.

On objective examination, periocular redness and swelling, mild ptosis of the left eye, as well as hyperemic bulbs and slight chemosis were noted laterally. There was no tenderness upon palpation. The ophthalmoscopy and pupillary responses were normal. A provisional diagnosis of preseptal cellulitis was made.

Because of the atypical symptoms, the patient was seen by ENT doctors who found normal flexible laryngoscopy. A computed tomography (CT) of the sinuses was also performed to rule out pre/postseptal cellulitis.

CT did not detect any signs of a subperiosteal abscess. However, inflammatory changes in the left lateral rectus muscle (Figs. 1–3), soft tissue swelling preseptal medially in the left lamina papyracea, and osteomeatal complex occlusion on the left side due to slight mucosal thickening in the sinus sphenoidalis/ethmoidalis were observed.

Due to some degree of sinusitis and suspicion of periocular infection, the patient was administered peroral antibiotic treatment with amoxicillin-clavulanate and discharged for outpatient follow-up.

After 2 days, the patient visited the ophthalmology department because of deterioration in the form of diminished vision and fading color vision in the left eye.

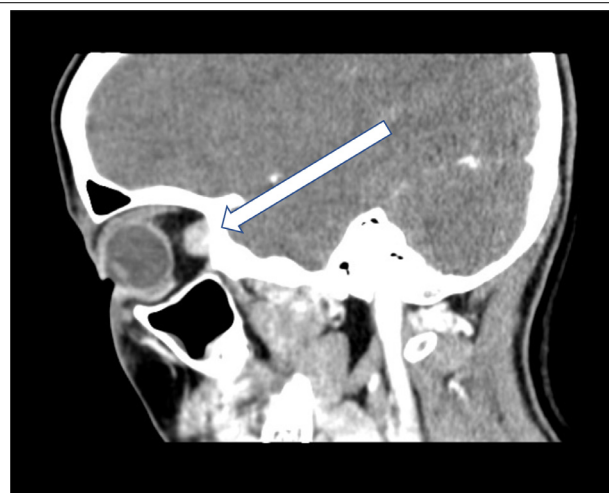


Fig. 3 – Contrast-enhanced CT scan, sagittal view, shows enlarged left lateral rectus muscle with contrast enhancement (arrow).

On objective examination, mild palpation tenderness, restricted abduction-adduction, affected upward eye movement, proptosis, universal chemosis and eye's hyperemia on temporal side were found. In addition, were observed decreased visual acuity (0.63) and color saturation despite normal fundoscopy.

Paraclinical showed normal infection parameters.

She was hospitalized, and treatment with intravenous antibiotics was initiated without appreciable effect.

The magnetic resonance imaging (MRI) showed a thickened left lateral rectus muscle at full length (Figs. 4–6), including the tendon attachment with increased contrast enhance-

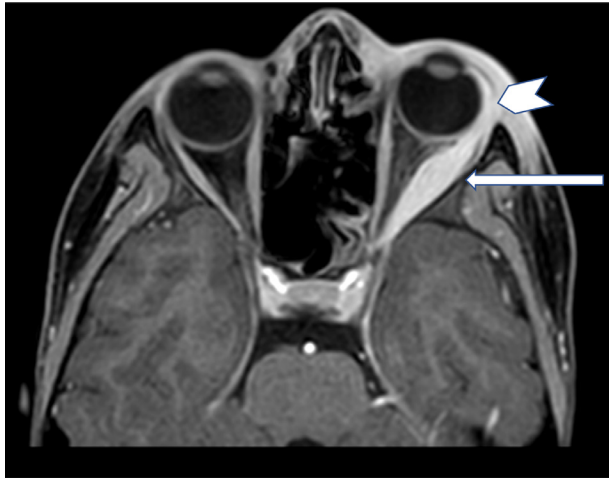


Fig. 4 – Contrast-enhanced MRI, T1, axial view shows enlarged left lateral rectus muscle with contrast enhancement (arrow) and attachment involvement (arrow head).



Fig. 5 – MRI, without contrast, T2, axial view shows enlarged left lateral rectus muscle (arrow) and attachment involvement (arrow head).

ment associated with inflammatory changes in the adjacent left preseptal soft tissues and left lacrimal gland.

Based on the clinical and radiological findings, the most likely diagnosis was myositis with concomitant orbital inflammation, due to which peroral steroid treatment was started, as well as administration of the antibiotics was continued.

A day after starting treatment, marked improvement was noted.

Whole-body MRI was performed to rule out rheumatological disorders or involvement of myositis in other muscles. MRI

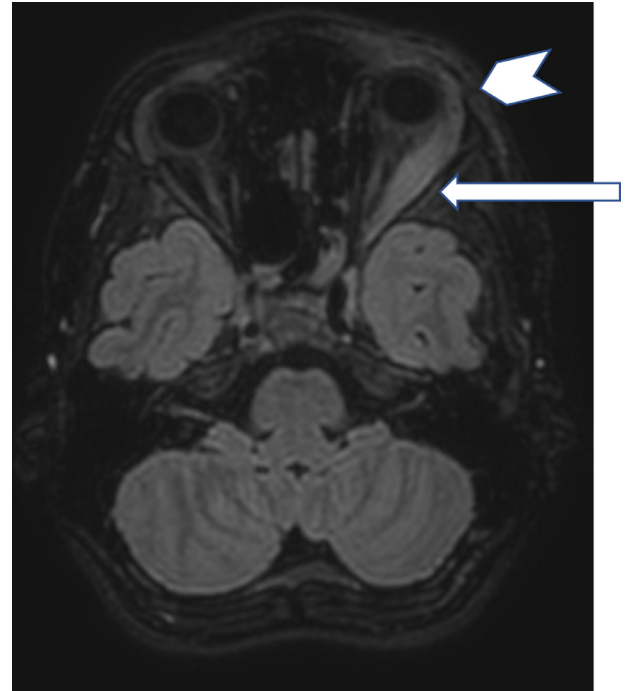


Fig. 6 – MRI, without contrast, T2 FLAIR, axial view shows enlarged left lateral rectus muscle (arrow) and attachment involvement (arrow head).

showed unchanged thickening of the left lateral rectus muscle, with no changes elsewhere.

Blood tests showed normal rheumatologic and endocrinologic parameters.

Steroids were slowly tapered over the course of the weeks. One month after the start of treatment, there were no complaints and a normal objective examination was performed.

Discussion

Idiopathic orbital inflammatory syndrome, also known as orbital pseudotumor, first described by Gleason in 1903 [3], is a benign nongranulomatous, noninfective, and non-neoplastic inflammatory process in the orbit in which a local or systemic cause cannot be established [4].

Orbital myositis is a subtype of IOI that primarily affects extraocular muscles [5]. The most affected muscles are the superior rectus, lateral rectus, and medial rectus muscles [6]. Other orbital structures may also be involved in this inflammatory process, including the lacrimal glands, orbital fat, sclera, uvea, superior orbital fissure, cavernous sinus, and the optic nerve. Extraorbital extension and bilateral eye involvement have also been reported [5].

IOI is usually seen in their fifth decade of life, and there is no sex predilection. However, orbital myositis most commonly affects young adults in the third to fourth decade of life and shows a female predilection [7].

The classical presentation is usually a clinical triad of peri-orbital pain, especially with eye movement, ophthalmoparesis, and signs of inflammation such as redness, chemosis, proptosis, and periorbital edema. Other clinical features include diplopia, restricted motion, ptosis, and possible involvement of the optic nerve, which, if left untreated, can lead to blindness [4]. As IOI is a diagnosis of exclusion, physical examination and clinical history are essential to rule out associated diseases, including systemic immune-related diseases such as orbital cellulitis, optic neuritis, thyroid ophthalmopathy, sarcoidosis, histiocytosis, Wegener's granulomatosis, Tolosa-Hunt syndrome, optic gliomas, lymphomas, and other neoplastic conditions. Infection is a common cause of IOI, which can be hidden in structures around the orbit [5].

In the emergency department, CT is the modality of choice for the diagnosis of orbital myositis. The typical findings on CT are homogeneous, fusiform enlargement of the muscle belly with enhancement, extending inferiorly to involve tendon insertion on the globe. However, the absence of tendon involvement does not rule out idiopathic orbital myositis [4,8–10].

The best imaging modality for idiopathic orbital inflammatory disease is contrast-enhanced thin-section MRI. Overall, the radiographic features of idiopathic orbital inflammatory syndrome vary widely. The best diagnostic clue is poorly marginated, mass-like, enhancing soft tissue involving any area of the orbit. Radiological findings include inflammation of the extraocular muscles (myositis) with tendinous involvement; orbital fat stranding; lacrimal gland inflammation and enlargement (dacryoadenitis); involvement of the optic sheath complex, uvea, and sclera; a focal intraorbital mass; or even diffuse orbital involvement. Though bone destruction and intracranial extension are rare, they have been reported [3,9,10].

For mild diseases, observation is acceptable with or without nonsteroidal anti-inflammatory drugs, such as ibuprofen. For moderate-to-severe disease, systemic steroids with slow tapering remain the mainstay of treatment. Chemotherapy with methotrexate may be considered in patients who are refractory or non-tolerant to steroids. The response to radiation therapy has also been documented [5].

Conclusion

Idiopathic orbital myositis should be considered in the differential diagnosis of orbital pain, especially that which is associated with eye movement, even in preteens. Diagnos-

tic imaging is very useful, particularly when other clinical signs of inflammation are missing. Systemic steroid therapy induces significant clinical improvement within a few days of treatment.

Patient consent

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

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