Idiopathic inferior rectus myositis: A rare cause of diplopia and a therapeutic success

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Abstract:
Orbital myositis is a rare inflammatory condition of a single or multiple extraocular muscles. It usually presents with unilateral or sequential bilateral sub-acute eye swelling, orbital pain, restricted eye movement, and redness of the eyelid. The swelling of extraocular muscles may lead to limited range of motion that produces blurred vision or diplopia[2]. The diagnosis of idiopathic orbital myositis is made after exclusion of other condition that may lead to orbital inflammation such as Grave’s disease, sarcoidosis, and malignancy[3]. Approximately 10% of patients have a concurrent autoimmune disease, which suggests that the dysfunction of the immune system may be a part of the underlying etiology[4]. In this article, we report a male patient with idiopathic inferior rectus myositis presenting with diplopia who was managed successfully by medical and surgical intervention. Acute onset of diplopia with eye pain or limited extraocular movement is an ophthalmic emergency requiring urgent assessment and diagnostic imaging studies such as CT or MRI. The present case shows the crucial role of surgery as an adjunctive modality to achieve an improved clinical picture in patients not responding to immunosuppressive therapy. The secret to the success of management includes regular follow-up with frequent examination and comprehensive radiological and tissue investigations.

Keywords:
Diplopia, inferior rectus, idiopathic, surgery, orbital myositis

INTRODUCTION
Orbital myositis is a rare inflammatory condition of a single or multiple extraocular muscles, formerly termed orbital pseudotumor[1]. It usually presents with unilateral or sequential bilateral sub-acute eye swelling, orbital pain, restricted eye movement, and redness of the eyelid. The swelling of extraocular muscles may lead to limited range of motion that produces blurred vision or diplopia[2]. The diagnosis of idiopathic orbital myositis is made after exclusion of other condition that may lead to orbital inflammation such as Grave’s disease, sarcoidosis, and malignancy[3]. Approximately 10% of patients have a concurrent autoimmune disease, which suggests that the dysfunction of the immune system may be a part of the underlying etiology[4]. In this article, we report a male patient with idiopathic inferior rectus myositis presenting with diplopia who was managed successfully by medical and surgical intervention.

CASE REPORT
A 73-year-old male presented to the neurology clinic with a 4-month history of binocular double vision, which increased when he looked downward. He had a history of hypertension, diabetes, gout, and hepatitis C carrier. There was no history of trauma, systemic malignancy, or thyroid disease. He denied any distant or near vision complaint, eye pain, headache, conjunctival redness. His medications included metformin, allopurinol, amlodipine, and atorvastatin.

Clinical examination revealed normal vital signs and general examination. Marked restriction of right eye elevation beyond primary position especially in dextroelevation (eye movement up and to the right). He had excyclotorsion in the right eye approximately 15 degrees. Forced ductions were positive in elevation. It was also associated with significant extorsion approximately 15 degrees. From primary position upward, he had diplopia which is made worse while his right eye is in dextroelevation position. Pupillary examination, visual acuity, corneal reflex, and funduscopy were normal with...
no proptosis. Other cranial nerve examination was normal, and the rest of neurological examination was normal including motor, sensory, cognitive, cerebellar, and gait.

Detailed investigations including basic biochemistry, thyroid function tests, thyroid antibodies, anti-acetylcholine antibody, anti-musk antibody, vasculitis and connective tissue screen, and tumor markers were all unremarkable. Computed tomography (CT) scan and magnetic resonance imaging (MRI) of the brain were unremarkable apart from evidence of minor small vessel disease. Orbital CT scan with contrast showed homogeneous elliptic shaped enlargement of the right inferior rectus muscle, which faintly enhanced post contrast injection. It measures 1.6 × 1.1 × 0.7 cm in anteroposterior, transverse and craniocaudal dimension. In addition, there was minimal proptosis [Figure 1]. Orbital MRI showed diffuse uniform enlargement of the right inferior rectus. The remaining orbital anatomical structures showed normal signal characteristics [Figure 2]. CT chest, abdomen, pelvis and bone scan were all unremarkable. A biopsy of the enlarged right inferior rectus muscle was performed and showed fibrovascular adipose tissue and smooth muscle with no inflammatory or malignant changes. An isolated monocular inferior rectus myositis was diagnosed according to the clinical, radiological, and histopathological findings with the exclusion of any other possible etiology.

The patient was treated with intravenous pulse steroid therapy (methylprednisolone 1 g daily for 5 days) followed by oral prednisolone (1 mg/kg/day for 1 month) in gradually tapering doses. Oral prednisolone was gradually and slowly tapered over a period of 6 months. Unfortunately, his response to steroid treatment was very minimal. Given the age of the patient and co-morbid conditions, aggressive immunosuppressive therapy such as methotrexate was rejected by the patient. His medical condition was stable, but his double vision did not improve. Six months after the onset of symptoms, the surgical option was explained to him, and he underwent right orbital decompression with right inferior rectus recession and hang-back from 7 mm to approximately 15 mm from limbus with lysis of intramuscular adhesions. His symptoms improved dramatically after surgery although he continued to see the double vision when he looked upward [Figure 3].

The patient stayed in good condition for the past 18 months, and he is still following-up regularly in our hospital and kept on a small dose of oral prednisolone (10 mg once daily).

Discussion

Orbital myositis is an idiopathic inflammation of the extraocular muscles with a 2:1 female predominance. It is often included under the broad spectrum description of pseudotumor (currently termed idiopathic orbital inflammatory syndrome)[5]. The condition was first described by Gleason in 1903[6]. The exact pathogenesis of orbital myositis is not fully elucidated. However, an immune-mediated mechanism with both B and T lymphocytes being involved remains the most acceptable theory. Complement-mediated microangiopathy may underlie the pathophysiology of orbital myositis. Molecular mimicry has been suggested to explain cases of orbital myositis that follow acute infections or systemic disease[7].

The clinical onset of orbital myositis may be acute, sub-acute, or chronic. The classical presentation is typically by orbital pain and double vision. The pain may be mild or severe that lead to sleep disturbances and is usually exacerbated by attempted eye movement. The absence of pain does not preclude the diagnosis of orbital myositis. However, it certainly raises concern for a neoplastic process, which would require a biopsy. On examination, patients typically show restriction of eye movements, edema and redness of the eyelids and conjunctiva, proptosis (which is generally minimal), and strabismus[8].

Several medical conditions can lead to extraocular muscle enlargement and demonstrate symptoms and signs of orbital myositis. These include thyroid, vascular, inflammatory,
neoplastic, neuromuscular, infectious, and metabolic disorders. The most common cause of extraocular muscle enlargement is by far thyroid disease. Approximately 80% of patients with idiopathic orbital myositis have a unilateral involvement.

The diagnosis requires the exclusion of other diseases including thyroid-associated orbitopathy, autoimmune myositis associated with systemic disease, carotid-cavernous fistula, and IgG4-related disease. Workup should include a full ophthalmic examination including assessment of optic nerve functions, extraocular motility, and exophthalmometry. Laboratory workup should include a thyroid function test, erythrocyte sedimentation rate, C-reactive protein, IgG4, ANCA, angiotensin-converting enzyme levels, and connective tissue screen.

Diagnosis of orbital myositis is confirmed by radiological evidence of enlargement and enhancement of one or more extraocular muscles, with or without the involvement of the myotendinous insertion. The tendon is spared in a muscle enlarged secondary to Graves orbitopathy. MRI of the orbit with/without contrast and with fat suppression is the best modality to evaluate this condition. CT of the orbits is a useful alternative if MRI is not readily available or when there is a clinical concern for an abscess or involvement of a bony structure of the orbits and adjacent sinuses.

Tissue biopsy obtained by a minimally invasive approach may be of importance in demonstrating the possible cause of orbital inflammation. The decision of biopsy should be guided by patient demographics, comorbidities, review of system, laboratory results, and radiological findings. Biopsy is urged in patients presenting in an atypical fashion. Histopathological analysis typically shows a nonspecific inflammatory infiltrate associated with granulomatous inflammation and sclerosis.

Management of mild cases of orbital myositis involves the use of high-dose nonsteroidal anti-inflammatory drugs. Many of these patients achieve good control of symptoms with the resolution of acute inflammation. Those patients may never require additional treatment. Systemic corticosteroids have been the mainstay treatment of this disorder especially in moderate to severe cases or in patients with multiple muscle involvement. The starting dose is typically 1 mg/kg/day with a rapid clinical response supporting the diagnosis of orbital myositis (evident in the first 12–24 h). Although some tumors can favorably respond to corticosteroid for a short time, the response is rarely as rapid and complete as orbital myositis. A gradual steroid taper (over 3–4 weeks) should follow the clinical response after adequate dosage and enough duration. If there is no response to steroids, failure of a taper, prolonged steroid use, or intolerable steroid-related side effects, immunomodulatory agents (e.g., cyclophosphamide, methotrexate, azathioprine, and cyclosporine) can be used. Focal radiotherapy and periorbital steroid injection may be used in refractory cases.

Patients with orbital myositis who do not respond to medical therapy may be offered surgical decompression. The procedure was first described by Dollinger in 1911. The goals of surgical intervention are to reduce or reverse vision loss, relieve orbital congestion, and prevent ocular surface damage. The complication rate of orbital decompression is less than 10% with the main complications being periorbital ecchymosis and edema, postoperative hemorrhage, and infection. More than 25% of patients report improvement in diplopia after surgery.

In the literature, 50–55% of patients with orbital myositis may experience a recurrence of symptoms requiring corticosteroids use. Bad prognostic factors include multiple muscle involvement, male gender, proptosis, and lack of response to systemic corticosteroids.

In conclusion, idiopathic orbital myositis is a rare inflammatory disease and the third most common cause of orbital inflammation (after thyroid orbitopathy and lymphoproliferative disease). Acute onset of diplopia with eye pain or limited extraocular movement is an ophthalmic emergency requiring urgent assessment and diagnostic imaging studies such as CT scan or MRI. The present case shows the crucial role of surgery as an adjunctive modality to achieve an improved clinical picture in patients not responding to immunosuppressive therapy. Diagnosis, assessment, and management of orbital myositis require a multi-disciplinary team including an ophthalmologist, a neurologist, and a clinical pharmacist. The secret to the success of management includes regular follow-up with

Figure 3: Postoperative images of our patient looking in different directions as indicated by the arrow in each image (b-i) with the exception of (a) where the patient is looking in the primary gaze position.
frequent examination and comprehensive radiological and tissue investigations.

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**Conflicts of interest**

There are no conflicts of interest.

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