Orbital Myositis

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Abstract

We report five cases of presumed orbital myositis mimicking extraocular muscle motility disturbances and manifesting clinical signs of active inflammation over the involved muscles. Computed tomographic evidence for extraocular muscle enlargement is helpful in confirming the diagnosis. If not present or atypical, another etiology should be sought. All patients responded rapidly and dramatically to systemic corticosteroids. Anterior inflammation may be accompanied by iritis and respond to topical corticosteroids. We believe the diagnosis of orbital myositis may be made on clinical grounds with confirmation by computed tomographic evidence for extraocular muscle enlargement and clinical response to corticosteroids. Biopsy is unnecessary except in atypical cases.

Introduction

The diagnosis of orbital myositis implies orbital inflammation confined to one or two extraocular muscles and may represent a distinct clinical entity mimicking a paretic extraocular muscle of acute onset, with restricted ductions. It is responsive to systemic corticosteroids.1-2 We report five patients with acute extraocular motility dysfunction presumably due to orbital myositis. The diagnosis, based upon suspicion, clinical presentation, and computed tomographic evidence, was confirmed by rapid resolution after treatment with systemic corticosteroids.

Case 1

A 37-year-old woman presented with a swollen, painful right eye and horizontal diplopia. Her symptoms had increased for 3 weeks prior to referral. She denied any antecedent upper respiratory infection or history of sinusitis. Examination revealed 20/15 acuity in each eye. The right eye was restricted in both abduction and adduction (Fig. 1). The conjunctiva over the medial rectus was inflamed (Fig. 2). Six millimeters of proptosis were present in the right eye. Slit lamp examination and intraocular pressures were normal as was funduscopic. B-scan ultrasonography revealed a markedly enlarged medial rectus, better demonstrated by computed tomography (Fig. 3a). There was no other evidence for orbital pathology or involvement of the adjacent sinuses (Fig. 3b). She was treated with 80 mg prednisone daily with a rapid resolution of both pain and proptosis. Steroids were tapered over 8 weeks. Three weeks later, while taking 40 mg prednisone per day, she was asymptomatic except for mild restriction of abduction of the right eye. Examination was otherwise negative. Two weeks later, while taking prednisone 10 mg per day, there was still limitation of ocular abduction. Eight weeks after presentation, she was asymptomatic and extraocular motility was normal. Prednisone was discontinued and she has remained asymptomatic for the past 6 months.

Case 2

A 16-year-old boy was referred with a diagnosis of orbital cellulitis. He related a 2-day history of pain and swelling of the right periorbital region. Afferent visual function was normal. Examination revealed 4 mm of right proptosis, swelling, and erythema of the right orbit, and mild limitation of abduction and adduction of the right eye (Fig. 4). Neuro-ophthalmologic examination was otherwise normal. B-scan ultrasonogram demonstrated enlargement of the right medial rectus. Computed tomography (Figs. 5a and 5b) demonstrated an enlarged medial rectus and a diffuse right pansinusitis involving the right frontal, ethmoid, and maxillary sinuses. He was treated with 80 mg of prednisone daily with resolution of orbital and sinus abnormalities over 48 hours. Steroids were rapidly tapered and decreased over a 2-week period. He has remained asymptomatic. Computed tomography 4 months later documented normal orbits and paranasal sinuses.

Case 3

A 53-year-old woman was referred with a 1-week history of pain in the left eye increasing with abduction. She was treated by her family physician with intramuscular penicillin and oral erythromycin for a "blocked tear duct." When she failed to improve, she was referred for evaluation. No other significant history was elicited. Visual acuity was
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Figure 1. Restriction of both ocular abduction and adduction due to enlarged, myositic, right medial rectus.

Figure 2. Localized conjunctival inflammation overlying medial rectus acutely inflamed by orbital myositis.

unimpaired. There were no localizing pupillary signs. The left orbit was exquisitely tender to palpation. A partial left ptosis was present. Extraocular motility was restricted in elevation and abduction (Fig. 6). There was obvious inflammation over the superomedial portion of her left globe. Two millimeters of left proptosis were present. Slit lamp

Figure 3a. Computed tomogram demonstrating marked enlargement of right medial rectus.

Figure 3b. Coronal reconstruction demonstrating isolated involvement of right medial rectus and absence of adjacent sinus involvement.

examination, intraocular pressures and funduscoppy were normal.

B-scan ultrasonography revealed a mass in the superomedial orbit. Computed tomography demonstrated a well-circumscribed mass in the superomedial orbit adjacent to and contiguous with the medial rectus (Fig. 7a); after injection of contrast, ring enhancement was evident (Fig. 7b). There was no computed tomographic or x-ray evidence for bony destruction or involvement of the adjacent sinus.

She was treated with 80 mg prednisone per day. Within 24 hours she was pain-free; after 72 hours she was asymptomatic. Prednisone was decreased
to 40 mg per day. Two weeks later, she was totally asymptomatic and her ophthalmic examination was normal. B-scan ultrasonography revealed the acoustically homogeneous medial orbital mass to be markedly decreased in size. Prednisone was rapidly tapered and discontinued over a 6-week period. She remained asymptomatic. A repeat computed tomography was obtained 6 weeks after her initial scan and demonstrated a marked reduc-

Figure 4. Periorbital swelling and mild limitation of ocular abduction and adduction, right eye.

Figures 5a and 5b. Computed tomograms (axial and coronal reconstruction) demonstrating enlargement of the right medial rectus and adjacent ethmoid sinusitis.
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of the globe was inflamed. Forced ductions were positive in the right eye confirming the presence of a pseudo-Brown's syndrome. The remainder of the ocular and orbital exam was normal. There was no x-ray evidence for sinusitis. A diagnosis of superior oblique myositis was made. She was treated with 80 mg prednisone per day with rapid resolution of her symptoms. Four days later, examination was normal. Steroids were rapidly tapered over 2 weeks. She was asymptomatic for 7 months. She then presented with mild iritis and a recurrent pseudo-Brown's syndrome. Both responded to topical 1% prednisolone acetate over a 2-week period. Three months later, her superior oblique myositis recurred accompanied by superior periorbital edema. Orbital computed tomography was negative, except for soft tissue swelling in the vicinity of the trochlea. She again responded to topical prednisolone acetate and is presently asymptomatic on a maintenance dose of topical prednisolone acetate twice daily.

Case 5

A 10-year-old girl was referred for evaluation of ptosis. Three weeks prior to evaluation, she awoke with a ptotic upper lid. Over the next 2 weeks, mild swelling and tenderness developed. After a negative laboratory and neuroradiologic evaluation, she was hospitalized and treated with intravenous ampicillin for presumed orbital cellulitis. When the swelling and erythema increased, she was referred for neuro-ophthalmologic consultation.

No significant additional history was elicited. Visual acuity and color vision were equal and normal. The left upper lid was ptotic with no levator function evident. The lid was mildly erythematous and swollen with a palpable mass distending the orbital septum. A left hypotropia was present and left superior rectus dysfunction was evident (Fig. 10). Two millimeters of left proptosis were present. The superior conjunctiva was inflamed. The remainder of the anterior segment and fundus examination was normal. B-scan ultrasonography demonstrated enlargement of the left superior rectus. X-rays of the sinuses were normal. Computed tomography, obtained prior to referral, demonstrated soft tissue swelling in the anterior and superior orbit. She had not been febrile and her white blood count was normal. After discontinuing the ampicillin, she was treated with 60 mg of prednisone daily. In 24 hours, her ptosis and superior rectus paresis had partially resolved as did her subjective symptoms. One week later, she was asymptomatic except for mild residual left ptosis. Extraocular motility was normal. Steroids were tapered and discontinued over 3 weeks. She has remained asymptomatic.
Figure 7a. Computed tomogram demonstrating well-circumscribed mass in supermedial orbit.

Figure 7b. Ring enhancement of orbital mass after injection of intravenous contrast.

Discussion

The first two cases represent orbital myositis confined to the medial rectus. The first patient had neither an adjacent sinusitis or an antecedent upper respiratory infection as described previously by others.3 The second patient demonstrated orbital myositis of the medial rectus associated with a diffuse pansinusitis, mimicking orbital cellulitis. Eshaghian and Anderson4 reported inflammatory orbital masses with adjacent sinus involvement mimicking malignant neoplasia of the sinus, invading the orbit through apparent erosion of the medial orbital wall. No such pseudoerosion was present in our patient. Our patient was referred with the diagnosis of orbital cellulitis. Since he was only mildly febrile (100°F) and had a normal white blood count, we suspected inflammatory orbital disease. Computed tomographic evidence of an enlarged medial rectus confirmed our diagnosis. Both these patients had
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Figure 8a. Follow-up computed tomogram demonstrating decreased size of orbital mass and loss of ring enhancement.

Figure 8b. Coronal reconstruction demonstrating mass to be superior to and adjacent to the left medial rectus.

the rapid and dramatic response to systemic corticosteroids reported by others. They also demonstrated that medial rectus myositis may occur with or without adjacent sinus involvement and without a significant antecedent upper respiratory infection. Follow-up computed tomography (case 2) documents total resolution of the inflammatory process in both the orbit and sinuses.

Our third case represented what we believe to be a variant of orbital myositis. She presented with a painful ophthalmoplegia thought secondary to orbital myositis and her symptoms responded dramatically to systemic corticosteroids within 24 hours. However, her computed tomography (Figs. 7a and 7b) was atypical, demonstrating an enhancing medial orbital mass that failed to resolve totally on repeat scan 6 weeks later (Figs. 8a and 8b).

Evaluation of coronal reconstructions obtained during her follow-up scan revealed the mass to be contiguous with the medial rectus (Fig. 8b). These scans are obviously different from those demonstrating obvious medial rectus enlargement. It has been suggested that this picture may represent the lymphocytic variant of idiopathic orbital inflammation. Shortly after the second computed tomography, extensive metastatic melanoma from a cutaneous primary was documented.

Although we have no histopathologic evidence, we wonder whether this orbital mass may also represent metastatic melanoma with a surrounding inflammatory response which responded dramatically to systemic corticosteroids. A discrete metastasis to an extraocular muscle from silent small cell carcinoma has been recently described. Although mimicking orbital myositis, this patient did not respond to systemic corticosteroids. However, our patient's orbital symptoms did not recur after discontinuing the corticosteroids. A follow-up scan was negative, suggesting an atypical form of orbital
myositis. We urge clinicians to be suspicious of the diagnosis of orbital myositis if the computed tomography picture is atypical in spite of a good clinical response to systemic corticosteroids, or if the presumed myositis fails to respond to corticosteroids.

Case 4 represented orbital myositis presenting as a pseudo-Brown's syndrome. Again, external signs of inflammation were present over the involved area. Computed tomography failed to demonstrate extraocular muscle enlargement, but we suspected a trochleitis or myositis based upon clinical findings (point tenderness, restriction of motion, and overlying inflammation). A concomitant anterior uveitis has been reported previously to accompany orbital inflammation7 and might be expected with an anterior site of inflammation. This patient also demonstrated that idiopathic orbital inflammation, if anterior, may be controlled with topical corticosteroids.

Case 5 represented involvement of levator and superior rectus by orbital inflammation in a child. Again, there was evidence for active inflammation (warmth, erythema, a palpable tender mass over the involved lid, and injection of vessels over the involved superior rectus). Computed tomography was nonspecific as obtained on an outmoded unit, demonstrating only swelling of the lid, compatible with a preseptal cellulitis. The diagnosis here was made on clinical grounds—levator and superior rectus dysfunction accompanied by evidence of inflammation and confirmed by the rapid response to corticosteroids. High-resolution computed tomography with appropriate coronal reconstructions might well have documented enlargement of the levator and/or superior rectus.

Orbital myositis represents a distinct clinical entity that may mimic many disorders of extraocular motility. Evidence for local orbital inflammation should obviate against a neurologic etiology for these motility disturbances. Orbital myositis must be differentiated from other causes of orbital inflammation with motility disturbances. We believe this can be done by clinical examination, laboratory data, and computed tomography.

The most important entity to differentiate is orbital cellulitis. In our experience, these patients are febrile greater than 102°F, have a polymorphonuclear leukocytosis greater than 15,000, exhibit extraocular motility dysfunction, and often have a concomitant sinusitis or history of trauma. Our patients with orbital myositis have been otherwise well, essentially afebrile, and have normal white blood cell counts.

Partially treated orbital cellulitis may be confused with orbital myositis. Case 5 was referred...
after her orbital swelling failed to respond to intravenous and then oral antibiotics. In this situation, without diagnostic, computed tomographic evidence, we suggest discontinuing antibiotics and observation for 24-48 hours. If the clinical status does not worsen or remains stable, the patient is treated with systemic corticosteroids expecting a prompt, dramatic response.

The patient with orbital myositis has an exquisitely tender orbit to palpation and an obvious motility defect often limited to one or two extraocular muscles. Vision has been unimpaired in all of our cases.

We believe the diagnosis of orbital myositis can be made on clinical grounds and confirmed by computed tomographic evidence of an enlarged extraocular muscle and rapid response to systemic corticosteroids. Clinical findings included limitation of extraocular motility in the field of action of the affected muscle or in the opposite field, coupled with evidence of overlying inflammation and exquisite tenderness to palpation. Proposis is often present, but may be minimal with more anterior lesions.

Computed tomography of the orbit may document markedly enlarged extraocular muscles thought to be distinguishable from enlargement due to Graves' orbitopathy. The tendon is spared in a muscle enlarged secondary to Graves' orbitopathy, whereas both muscle and tendon are enlarged (Figs. 3a, 3b, 5a, and 5b) when involved by myositis. Other causes of extraocular muscle enlargement on computed tomography such as metastatic tumors, carotid cavernous fistulae, and trauma should be discernable by history, examination, and failure to respond to corticosteroids. Concomitant sinusitis may also be documented by computed tomography (case 2, Figs. 5a and 5b).

We agree with Purcell and Taulbee that orbital myositis is a distinct clinical entity. However, we find no consistent relationships to antecedent viral or streptococcal upper respiratory infections. Adjacent sinusitis may or may not be present. When present (case 2), it responds as dramatically as the orbital involvement to systemic corticosteroids. Eshaghian and Anderson have demonstrated similar histopathology in idiopathic orbital and adjacent sinus inflammation.

We do not postulate an etiology for orbital myositis but urge its recognition as a diagnostic entity by the clinician. Such recognition obviates the need for biopsying these inflamed orbits with their higher incidence of postoperative complications. We also recognize that some tumors can respond favorably to corticosteroids for a short time, but rarely as rapidly and completely as orbital myositis. We urge biopsy of patients presenting in an atypical fashion, possibly by fine-needle aspiration under sonographic guidance as described previously.

Patients with orbital myositis may be extremely uncomfortable and, in a referral practice, far from home. Their true clinical picture may be obscured by previous antibiotic treatment or surgery. Accordingly, our treatment regimen consists of hospitalization for observation and to expedite obtaining computed tomography. Treatment consists of 50 mg prednisone on admission followed by 80 mg daily in four divided doses. A dramatic subjective and objective improvement is usually evident in the first 12-24 hours. Corticosteroids are rapidly tapered over 3-4 weeks. Recurrences may be treated again with systemic corticosteroids. One patient (case 4) is successfully maintained on low-dose topical steroids after refusing further systemic prednisone. Patients who fail to respond to this regimen, or have an atypical clinical picture, subsequently undergo either open or fine-needle aspiration biopsies as their clinical condition dictates.

In essence, orbital myositis is a clinical diagnosis based upon suspicion, signs of extraocular muscle dysfunction with concomitant overlying inflammation, and a rapid response to systemic corticosteroids. Computed tomographic evidence for enlarged extraocular muscles is helpful; however, if the computed tomographic picture is atypical, another etiology should be considered and a histopathologic diagnosis obtained.

References

5. Personal communications with S. Trokel.

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