Ocular Myositis: A Case Study

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ABSTRACT

Orbital myositis is an uncommon inflammatory condition resulting in variable degrees of restriction of the extraocular muscles. A case of a 15-year-old girl is presented, highlighting the importance of differential diagnosis from other ocular conditions that can cause extraocular muscle restrictions.

Keywords: orbital myositis, extraocular muscles, inflammation, restriction, diplopia

INTRODUCTION

Ocular myositis is an idiopathic inflammatory condition in which orbital inflammation is confined to the extraocular muscles and occurs in the absence of inflammation of other orbital or peri-orbital tissues.[1,2] It is classified as one of the patterns of clinical presentation of orbital pseudotumour, a condition distinguished by inflammation of any orbital soft tissue, including orbital fat, lacrimal gland and connective tissue.[3]

The condition is characterised by an acute onset of orbital pain, often exacerbated by eye movement, diplopia, proptosis, duction restrictions, ptosis and conjunctival injection. Visual acuity and optic nerve function remain intact in the presence of the extraocular muscle inflammation.[4,5]

Cases of ocular myositis may be acute or chronic in presentation. Acute or isolated cases of ocular myositis are those that present with a recent onset of symptoms, normally less than two weeks, including pain and/or diplopia. The more chronic cases of orbital myositis include episodes that continue for a period of more than two months, or recurrent acute episodes which can lead to long term extraocular muscle restriction. Atypical cases of orbital myositis have also been reported.[6] These cases include those with uncharacteristic presentations including lack of pain or optic nerve dysfunction.

The cause of ocular myositis at this stage is unknown but it is hypothesised that an immune-mediated process may be involved following reports of associations between systemic conditions (sarcoidosis and Crohn's disease) and the development of ocular myositis.[5,6]

This paper will present a case study of a 15-year-old girl diagnosed with chronic orbital myositis.

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CASE REPORT

In January 2010 a 15-year-old girl, Miss J, attended the Sydney Children's Hospital eye clinic following a three-week history of variable right ptosis and diplopia on down gaze. These symptoms were accompanied by a right-sided temporal headache and pain behind her right eye that had been present for the previous three months, with minimum relief from paracetamol.

Miss J had a history of similar symptoms with the exception of diplopia on two previous occasions. These episodes were investigated by an ophthalmologist and paediatrician respectively and no abnormality found on examination or on her computed tomography (CT) or medical resonance imaging (MRI) scans.

Interestingly, there was a strong family history of conditions affecting the extraocular muscles. Miss J's maternal aunt was diagnosed with myasthenia gravis and four male members of her family (both immediate and extended) were diagnosed with ocular myositis.

On initial observation Miss J displayed a right partial ptosis, which was confirmed by measurement of the palpebral aperture with the right being 7 mm and the left being 14 mm. Her visual acuity without glasses was right 6/6 and left 6/5. On cover test at 6 metres she demonstrated orthophoria. Cover test at 1/3 metre revealed a small

exophoria with an accommodative target. Cover test at 1/3 metre repeated with a non-accommodative target (a torch) showed decompensation of the latent deviation to a small right exotropia with a small right hypertropia. On measurement, this deviation was neutralised with 4 prism dioptres base-in with the vertical measurement varying between 14 and 18 prism dioptres base-down, fixing left, restoring binocularity. Binocularity was tested using the Lang Stereotest, on which Miss J demonstrated a positive response achieving 550 seconds of arc.

On testing ocular movements, there was marked restriction of movement of the right eye in most positions of gaze. There were -4 underactions of the right superior rectus, inferior rectus and superior oblique. There were -3 underactions of the right inferior oblique and a mild underaction of the right medial rectus of -1.

The patient was referred to the immunology and neurology departments for multidisciplinary review. All investigations by immunology and neurology departments were found to be unremarkable. She was also sent for CT of the orbits and CT angiogram to rule out possible cerebral aneurysm. CT of the orbits revealed marked enlargement of the right superior and medial rectus muscles with involvement of the myotendinous junctions.

Miss J was treated promptly with a high dose of oral prednisone, with 50 mg for three days, tapered to 25 mg for a further three days. A week after high-dose steroid treatment saw an improvement in the patient's signs and symptoms, with almost complete resolution of diplopia and minimal extraocular muscle restriction remaining two months later.

Over the following year, the patient's condition improved but did not completely resolve. She had two episodes of relapse in which there was deterioration and changes in the affected extraocular muscles. Following each episode, maximum improvement in signs and symptoms occurred three to four weeks after commencing treatment. Each recurrence of ocular myositis coincided with reductions in the patient's steroid dosage below 5 mg. As a result of the patient's condition deteriorating with reductions in steroid dose, she has remained on a constant low-dose of steroids since initial onset of her condition, with increases in dosage when she has a replapse episode. Due to the long-term use of steroids the patient commenced treatment with methotrexate (a steroid-sparing agent) to help reduce the side effects of long-term steroid use.

DISCUSSION

Ocular myositis is a distinct clinical entity however extraocular muscle enlargement is a clinical feature also seen in other conditions including thryoid orbitopathy, carotid cavernous fistulas, metastases and infiltrative conditions.[6] Careful investigation of clinical characteristics is necessary for correct differential diagnosis to be made. Modern ultrasound and radiological techniques, allow the differential diagnosis of enlargement of the extraocular muscles to be promptly narrowed to ocular myositis and thyroid orbitopathy.

Thyroid orbitopathy is reported as the most common cause of enlargement of the extraocular muscles.[5] For cases of non-thryoid-related extraocular muscle enlargement, inflammation (classified as idiopathic orbital inflammatory disease or orbital pseudotumour) has been reported as one of the most common causes of extraocular muscle enlargement,[5,6,7] followed by vascular and neoplastic causes.[6,7]

The presenting signs and symptoms of thyroid eye disease and ocular myositis differ despite both having characteristic enlargement of the extraocular muscles. Patients with thyroid orbitopathy often present with a gradual onset of bilateral problems, often asymmetric, including dry eye, irritation, proptosis, and diplopia. Patients with ocular myositis will present with a more acute onset of symptoms, often unilateral, including pain on or exacerbated by eye movement, swelling and diplopia. Characteristically, in ocular myositis inflammation is isolated to extraocular muscles, whilst in thyroid orbitopathy a characteristic increase in orbital fat, causing exophthalmos, occurs in conjunction with enlarged extraocular muscles.

Diagnostic imaging (computed tomography) allows for differentiation between the two conditions as there is a distinct difference in the pattern of extraocular muscle enlargement and muscle involvement. In thyroid orbitopathy, bilateral asymmetric multiple muscle involvement is observed with regular muscle enlargement confined to the muscle belly, sparing the tendinous insertions. The inferior rectus is the muscle most frequently involved, followed by the medial, superior and lateral recti muscles.[2,7] In contrast, the most common presentation of ocular myositis is unilateral with only a single muscle affected.[6] Ocular myositis can also present with multiple muscle involvement and it has been suggested that multiple muscle involvement at initial presentation may be a risk factor for recurrent episodes or chronic cases of the condition.[6] The horizontal recti muscles tend to be most commonly involved in cases of ocular myositis,[2,4] with vertical recti muscles and obliques less commonly involved.[2,8,9] Miss J's case of chronic ocular myositis which initially presented with unilateral enlargement of the medial rectus and superior rectus of the right eye, agrees with the literature in demonstrating cases of ocular myositis that initially present with multiple muscle involvement can be associated with recurrent episodes or chronic ocular myositis.

Diagnostic imaging performed on Miss J also revealed inflammation of the myotendinous junction in addition to enlargement of the extraocular muscles. The pattern of

muscle enlargement in ocular myositis seen on imaging tends to be irregular often with inflammation of the tendinous insertion on the globe. Although the 'tendon sign' has been identified as a reliable indicator of ocular myositis, with 40-53% of cases by Mannor et al,[4] and 70% of cases by Zulfiqar et al[2] reporting tendon involvement, an absence of tendon involvement does not disclude a diagnosis of ocular myositis.

A pathognomonic sign indicative of ocular myositis is a rapid and dramatic improvement in signs and symptoms once treatment has commenced with high doses of systemic corticosteroids.[4,6,10] The most notable improvement in symptoms occurs within a period of 3 to 5 days,[1,9,10] after which the high steroid dosage is tapered. Similar to Miss J, the majority of patients experience a complete resolution of signs and symptoms approximately one month following initial onset. However, as seen in our case study, tapering the dosage of systemic steroids has been reported to coincide with recurrent episodes of myositis.[10] Long-term systemic steroid use in some patients becomes intolerable and other treatment options require consideration. These include treatment with steroid-sparing agents (or adjuvant drugs) such as methotrexate,[6] which was used as an adjunctive therapy for Miss J due to her long-term steroid use, or more radical treatment with radiation therapy.[3,11] It has also been reported that initial stages of treatment with non-steroidal anti-inflammatory drugs is also an option, however this treatment option tends to lend itself to patients with a non-recurrent or acute ocular myositis.[4]

Although it is suggested that ocular myositis is an immune-mediated process, familial influence on the development of this condition may also be important to consider. Our patient reported four of her male family members had been diagnosed with ocular myositis. At this stage, it seems only one other study has reported a family with multiple members demonstrating symptoms suggestive of ocular myositis.[12] Therefore it could be hypothesised that genetic predisposition may play a part in the development of the ocular myositis. However, it is important to consider equally that other events may influence the expression of the condition even in the presence of genetically predisposing factors.[12]

CONCLUSION

Ocular myositis is characterised by enlargement of extraocular muscles visible on medical imaging and rapid improvement of symptoms following treatment with high dose systemic corticosteroids. Although it is a distinct clinical entity, thorough clinical investigation is required for this condition to be differentially diagnosed from other causes of enlarged extraocular muscles including thyroid eye disease, vascular disorders and neoplastic disease.

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