CLINICAL VIGNETTE

A Case of Orbital Myositis

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Case Report

A 32-year-old male presented to rheumatology for persistent orbital myositis. The patient had first developed orbital myositis 10 years prior when he developed sharp pain behind the eyes as well as double vision; left more so than the right eye. MRI of the orbits at that time indicated orbital myositis and he was treated with IV steroids followed by a prednisone taper with improvement. Over the next 2 years following diagnosis, he required 3 more steroid tapers due to recurrence of symptoms. After these episodes, his eye pain and double vision never returned to normal, but symptoms were minimal, so he did not seek further treatment.

Three months prior to presentation, he experienced a flare with worsening bilateral eye pain, eye swelling and double vision with worse symptoms in the left eye. He saw an ophthalmologist who ordered a MRI of the brain and orbits. The MRI showed enlargement of the right lateral rectus and left medial rectus muscles with restriction in abduction in the left medial rectus muscle. He then underwent a biopsy of the left medial rectus muscle that indicated chronic myositis. He was started on prednisone 60mg daily with improvement of symptoms. However, this time he was unable to taper prednisone below 40mg daily without recurrence of his symptoms.

Upon initial presentation to the clinic, the patient did not have any significant ocular pain but still experienced episodes of double vision. His main complaints were steroid induced insomnia, anxiety and weight gain. He had no other systemic symptoms. His past medical history was significant for having Attention Deficit Hyperactivity Disorder, ADHD. No prior surgeries other than the ocular muscle biopsy. His medications included prednisone 40mg daily for the myositis and amphetamine-dextroamphetamine 30mg daily for his ADHD. He does not smoke, denies any drug use and drinks alcohol socially. He is single without children and works as an artist. There is no known family history of autoimmune disease.

His physical exam was significant for left eye strabismus without ocular injection or orbital swelling. His heart and lung exams were normal. He had no rashes, joint or muscle pain. His neurologic exam was otherwise unremarkable with normal 5/5 strength in all 4 extremities both proximally and distally. Orbital myositis had already been confirmed by MRI and muscle biopsy. He was sent for additional lab testing in preparation for steroid sparing treatment.

Laboratory studies showed normal CBC, comprehensive metabolic panel, sedimentation rate, C-reactive protein, TSH, CK and aldolase. Infectious labs were negative for HIV, Hepatitis B and C, syphilis, TB and lyme. ACE levels were normal. Autoimmune serologies were negative for ANA, ANCA, dsDNA, SSA, SSB, Smith, RNP, centromere, Scl-70, anti-phospholipid antibodies, rheumatoid factor, CCP antibodies, TPO, thyroglobulin and Jo-1 antibodies. Thiopurine methyltransferase (TPMT), an enzyme that metabolizes thiopurines medications, was normal.

Given the normal TPMT enzyme, the patient was started on azathioprine 2mg/kg PO daily as a steroid sparing agent. His prednisone was tapered slowly off over a period of 6 months with success. He has maintained on azathioprine alone with stabilization of his orbital myositis.

Discussion

Orbital myositis is one subset of orbital inflammatory diseases; a general term encompassing inflammatory diseases that affect some or all of the structures contained within the orbit.[1] Orbital myositis is a rare inflammatory disorder that is localized to the extra-ocular muscles of the eye and represents a subgroup accounting for 8% of all idiopathic orbital inflammation.[2] Concurrent autoimmune disease has been shown to be present in approximately 10% of patients diagnosed with idiopathic orbital inflammation and most cases of orbital myositis occur in the absence of any form of chronic systemic illness.[3]

The classical presentation of orbital myositis is an acute to subacute onset of unilateral painful ophthalmoplegia associated with signs of ocular inflammation. The orbital pain typically worsens with eye movements and can be moderate to severe in intensity. The absence of orbital pain does not rule out orbital myositis as up to 50% of patients may not have pain. Multiple extra-ocular muscles may be affected leading to diplopia and a restrictive pattern of strabismus. Visual acuity is generally spared.[4] Unilateral presentation is typically seen but bilateral cases are not uncommon. Examination usually reveals restriction of eye movements and edema of the eyelids and conjunctiva. In severe cases, a mild proptosis may be observed due to retro-orbital inflammation.[2]

MRI is the imaging study of choice when orbital myositis is suspected.[4] The changes of orbital myositis are best seen on T2

fat suppressed post-gadolinium scans[2] and typically show thickening of the affected extra-ocular muscles involving the myo-tendinous junction and the surrounding fat with contrast enhancement.[4] Definitive diagnosis may require a biopsy of the muscle and surrounding orbital fat.[2] Although an orbital muscle biopsy might prove diagnostic in orbital myositis, it is sometimes contraindicated due to the risk of permanently damaging or compromising the function of the extra-ocular muscle.[3] The main utility of an orbital muscle biopsy is to rule out possibility of a neoplastic tissue or lymphoproliferative process and therefore can be indicated in treatment resistant cases such as this case or if imaging suggests the presence of an orbital tumor.[4]

The diagnosis of ocular myositis is considered one of exclusion, as there are many diseases that manifest with orbital inflammation.[3] A specific myositis associated with a systemic disease is a rare form of orbital inflammation but important to evaluate as treatment directed to the underlying cause would be required.[2] Autoimmune diseases that can be associated with ocular myositis include Grave's disease, sarcoidosis, systemic lupus, Crohn's disease, psoriatic arthritis, Behcets, ANCA-associated vasculitis and IgG4-related disease.[4] History, physical exam and lab testing should include assessment for these conditions as part of an evaluation in a patient with ocular myositis. In this case, there was no evidence of an underlying autoimmune disease as he had no extra-ocular symptoms and his autoimmune serologies were normal.

Since the main treatment of ocular myositis is steroids and immune suppression, infectious and malignant causes of inflammation must also be ruled out prior to initiating treatment.[1] The presence of fever, elevated white cell count and sinus disease on imaging should prompt consideration of an infectious cause.[2] Orbital cellulitis from staphylococcus or streptococcus species are the most likely causative organisms but cases of sino-orbital aspergillosis and rhino-orbital mucormycosis have also been reported.[1] Neoplastic processes, whether locally infiltrative or metastatic can also cause enlargement of extra-ocular muscles. Orbital lymphoma and lymphoproliferative diseases are the most common malignant causes, but metastatic lesions to the extra-ocular muscles have been reported from cutaneous melanoma, breast cancer and gastrointestinal malignancies.[2] This patient did not have any signs of infection and his ocular biopsy was negative for malignancy as well.

The mainstay of initial therapy in ocular myositis is oral corticosteroids.[2] Treatment with steroids often leads to rapid and robust relief of symptoms[3] which also supports the diagnosis of ocular inflammation. A failure to respond adequately to corticosteroids should trigger biopsy or further investigation prior to start of immunosuppressive drugs.[2] Even though inflammation tends to respond well to steroids, half of patients will relapse.[1] Azathioprine and methotrexate have been used successfully as steroid sparing agents. Case reports have also cited anti-TNF therapy as another possible effective treatment in resistant cases.[2] This patient responded well to azathioprine treatment and was able to taper off steroids without recurrence of ocular inflammation.

In conclusion, ocular myositis is a rare inflammatory condition that involves the extra-ocular muscles of the eye. Evaluation should include underlying autoimmune, infectious and malignant etiologies, but most cases occur without underlying systemic disease. Treatment with corticosteroids are effective but relapse rates are high and steroid sparing immunosuppressive therapy may be needed for better disease control.

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