CASE REPORT

A RARE CASE OF ORBITAL MYOSITIS

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The orbital myositis is a rare inflammatory disorder of extraocular muscles and is considered a subtype of nonspecific orbital inflammatory syndrome. It is usually a unilateral disease process with a limited course. We report a case of a 34-year-old man who was referred to the Radiology Department at Shifa International Hospital, Islamabad with 6 months history of severe pain, swelling, and redness in the left eye associated with blurring of vision and headache. Initially, the patient had contrast enhanced CT scan orbits which showed asymmetrically thickened, minimally enhancing left optic nerve having indistinct margins with thickened superior rectus and superior oblique muscles, indistinct superior ophthalmic vein, soft tissue nodularity in the medial intraconal space fat, and slight left exophthalmos. Differentials of orbital pseudo tumour and lymphoma were given. Further evaluation by post contrast MRI with orbits protocol was suggested which showed proptosis, the significant infiltrative disease in the superomedial aspect of left orbit involving the adjacent fat with thickening, enlargement, and heterogeneous enhancement of the superior rectus, medial rectus, and superior oblique muscles with relatively lesser degree involvement of their tendons. The infiltrative thickening also involved the optic nerve sheath and seen invading the optic nerve at the orbital apex. On basis of the imaging features and clinical picture, a diagnosis of orbital myositis with optic neuropathy was given. The patient was given corticosteroids and showed rapid symptomatic improvement.

Keywords: Orbital myositis; Inflammatory disorder; Extra-ocular muscles; Contrast enhanced MRI orbit; Corticosteroids

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INTRODUCTION

Orbital myositis is a rare inflammatory disease of the eye which is difficult to distinguish from other ocular pathologies that present with overlapping features. When autoimmune in origin, it presents with a positive autoimmune profile and shows a good response to corticosteroid therapy. Though it commonly presents in females around 32 years of age yet we present a case of a 34-year-old male presented with pain and diplopia, and her imaging features raised the suspicion of orbital myositis. Though the autoimmune profile turned out to be negative yet the patient responded to corticosteroid therapy and hence diagnosing was confirmed.

CASE PRESENTATION

A 34-year-old Caucasian man was referred to the Radiology department at Shifa international hospital, Islamabad with a 6 months history of severe pain and swelling in the left eye, associated with redness in the eye, blurring of vision, and headache.

Initially, the patient had CT scan orbits with and without contrast which showed asymmetrically thickened, minimally enhancing left optic nerve having indistinct margins with thickened superior rectus and superior oblique muscles, indistinct superior ophthalmic vein, soft tissue nodularity in the medial intraconal space fat, and slight left exophthalmos. Differentials of orbital pseudo tumour and lymphoma were given and further evaluation with post contrast MRI with the orbital protocol was suggested.

After a week, the patient had MRI orbit which showed significant thickening, and infiltrative disease in the superomedial aspect of left orbit involving the adjacent fat with thickening and disease infiltration resulting in enlargement and heterogeneous enhancement of the superior rectus, medial rectus, and superior oblique with relatively lesser degree involvement of their tendons. The infiltrative thickening also involved the optic nerve sheath and is invading into the optic nerve at the orbital apex with left sided proptosis. A suggestion of orbital myositis with optic neuropathy was given.

The patient was given corticosteroids. There was a symptomatic improvement. Hence the diagnosis of autoimmune orbital myositis was given.

DISCUSSION

The term orbital myositis refers to inflammation of extraocular muscles of eye, and accounts for about 8% of all the idiopathic orbital inflammations/orbital pseudotumor which is non-specific inflammation that can affect any structure in the orbit.[1] It can present at any age, commonly in the 4th decade of life with the incidence being more in females than males and male to female ratio of 1:2.[2] It is unilateral in 90% of cases but can be bilateral as well. The patient can have a variable clinical presentation from simple orbital discomfort to severe pain, diplopia on the movement of the eye, to some more severe forms having ptosis, chemosis, or proptosis.[3]

Though not very common in males in case it was a 34-year-old male who presented with unilateral eye pain and diplopia.

It can have an acute or chronic course, infectious or autoimmune in origin. When it is autoimmune in origin, it is usually associated with other autoimmune diseases.[4] Its presentation can mimic other causes of orbital inflammation like thyroid-associated orbitopathy, lymphoproliferative disorders, metastatic orbital diseases, myasthenia gravis, amyloid deposition, and infections.[5] But depending on clinical presentation and imaging features, these conditions can be distinguished from the other.

As in thyroid associated orbitopathy, the patient usually presents with painless bilateral asymmetrical as lid retraction, lid lag in downgaze, and abnormal thyroid function tests. On computed tomography scan it shows extraocular muscle enlargement involving muscle bellies and sparing tendons, giving a coca-cola bottle sign, whereas in orbital myositis patients presents with pain and imaging features which show involvement of tendons along with muscle bellies and normal thyroid function tests[6], which was typically present in our case.

Other differentials of primary or secondary malignancy of the eye can present with imaging features like that of orbital myositis but will have rarely have an acute and painful onset. Though not performed in our case biopsy can also be confusing as orbital myositis also shows lymphocytes which can be mistaken for orbital lymphoma, plasma cells, and giant cell infiltration.

Autoimmune orbital myositis usually does not have any direct association with other systemic disorders except for the underlying immune disorders such as linear scleroderma, SLE, rheumatoid arthritis, Crohn's disease, collagen vascular disorders, and allergy[7,8], in which case the patient had positive autoimmune profile specific for an associated autoimmune disease like Antinuclear Antibodies (ANA) Anti-DNA and Anti-Smith in SLE, Rheumatoid factor in RA, Anti-centromere in limited cutaneous scleroderma (CREST), Anti-acetylcholine receptor protein and Muscle-specific receptor tyrosine kinase (MuSK) antibody in Myasthenia Gravis. None was positive in our case.

Imaging features of orbital myositis include enlargement of the muscle belly of one or more extraocular muscles with the involvement of their tendinous insertion with or without inflammation of the surrounding tissues, including the lacrimal gland. Sometimes it can present as an infiltrative mass that extends outside of the orbit via superior or inferior orbital fissures, into cavernous sinus, meninges, and dura. (Figure-1).

MRI specifically shows T1 isointense to hypointense signals with corresponding T2 hypointense (in case of fibrosis), iso- to hyperintense otherwise. Postcontrast T1 C+ (Gadolinium) shows moderate to marked diffuse enhancement. (Figure-2).

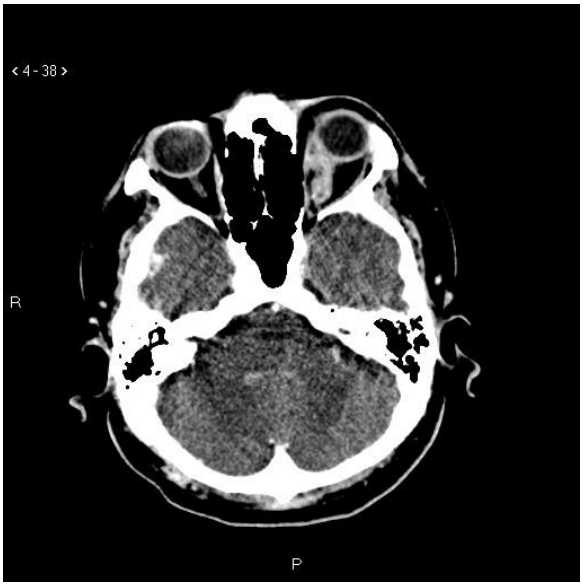

Figure-1: Axial CECT scan of brain at the level of orbit; showing thickened, enhancing left optic nerve with indistinct margins, thickened superior rectus and superior oblique muscles, indistinct superior ophthalmic vein, soft tissue nodularity in the medial intraconal space fat, and slight exophthalmos.

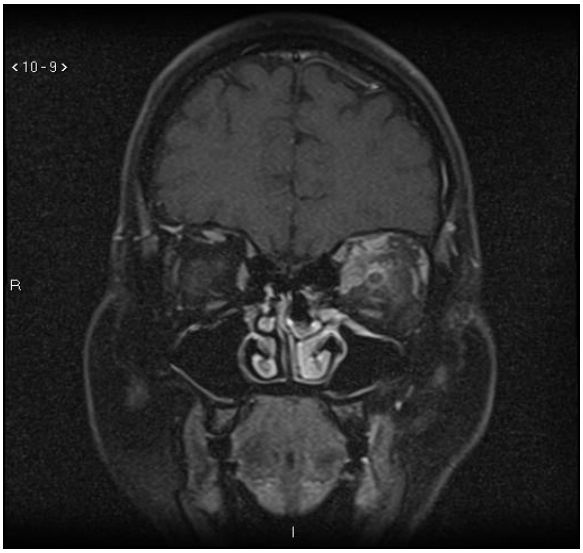

Figure-2: Coronal CE MRI of brain showing significant thickening and infiltrative disease in the superomedial aspect of left orbit involving the adjacent fat with resultant thickening of orbital muscles and associated enhancement along optic nerve.

CONCLUSIONS

Orbital myositis must always be taken into consideration in a patient with recurrent palsy of extraocular muscles, periorbital pain, and double vision. It is a diagnosis of exclusion and axial and coronal MRI of the orbit with fat suppression plays an important role in diagnosing this condition by accurate identification of involved muscles. The treatment includes corticosteroids and the degree of response gives a hint to the diagnosis.

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