Non-Specific Orbital Inflammation Extending Into The Cavernous Sinus : A Case Report

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Case Summary :

A 39 year old male patient presented to the ophthalmology out-patient department with progressive blurring of vision in his left eye for a period of 2 months associated with ocular pain, redness and swelling in the region of the left eye. On examination, there was left-sided periorbital fullness, proptosis, relative afferent pupillary defect (RAPD) and restriction of ocular movements in the left gaze by 10 degrees. Best corrected visual acuity of counting finger till 3 m on the left eye, and 6/6 on the right eye. Fundoscopy revealed marked disc edema. The neurological evaluation was unremarkable. Apart from screening blood tests, the patient underwent CT and MRI evaluation of the bilateral orbits. Biopsy of the fibro-adipose tissue of orbit revealed inflammatory cells consistent of inflammatory pseudotumor. High dose systemic steroid was instituted, followed by slow tapering of oral steroid. His symptoms completely resolved, and latest visual acuity was 6/9, with no recurrence, to date.

Introduction :

Non-specific orbital inflammation is a non-granulomatous inflammatory process in the orbit without any underlying local or systemic causes, presents with a variety of non-specific clinical symptoms, making diagnosis a challenge in majority of the cases. Conditions such as orbital neoplasm and orbital cellulitis always need to be excluded to eliminate possibilities of life threatening or other vision threatening diseases. Although, biopsy is the most ideal method of confirming the diagnosis, this may not always be amenable as the risks of injuring other orbital structures. Magnetic resonance imaging (MRI) has become the most important diagnostic tool in making the diagnosis. However, orbital cellulitis, orbital lymphoid lesion and orbital inflammatory syndromes are not readily differentiated in (MRI). This case-report is an attempt to unravel and shed some light on this rare presentation of non-specific orbital inflammation.

Case-Report :

A 39 year old male patient presented to the ophthalmology out-patient department with progressive blurring of vision in his left eye for a period of 2 months associated with ocular pain, redness and swelling in the region of the left eye. This was his first episode, and he was asymptomatic prior to this presentation. There were no constitutional symptoms reported. Patient does not have any history of diabetes mellitus or history suggestive of thyroid abnormality.

Best corrected visual acuity at the time of presentation was counting finger till 3 m on the left eye, and 6/6 on the right eye.

On examination, there was left-sided periorbital fullness [Fig. 1], proptosis [Fig. 2], relative afferent pupillary defect (RAPD) [Fig. 3] and restriction of ocular movements in the left gaze by 10 degrees. Rest anterior segment findings (OS) were unremarkable. Right eye had apparently normal anterior segment findings.

Imaging :

The computed tomography (CT) showed a well-delineated, enhancing, post-septal mass measuring approximately 2.7 cm x 1.2 cm in the left orbit along the left lateral rectus and superior rectus muscles. The mass was extending into the right cavernous sinus [Fig. 5].

On magnetic resonance imaging (MRI), the mass showed intense enhancement and involved the left lateral rectus and superior rectus. A soft-tissue enhancing mass

Fig. 1 : Left-sided periorbital fullness and mild conjunctival congestion

Fig. 2 : Left-sided proptosis

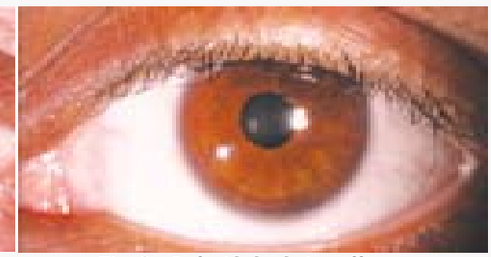
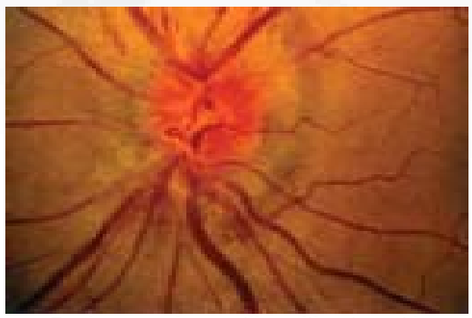


Fig. 3 : Left-sided relative afferent pupillary defect (RAPD)



Fundoscopy after pupillary dilatation with tropicamide 1%, revealed marked disc edema [Fig. 4].
Fig. 4 : Marked disc oedema (OS)

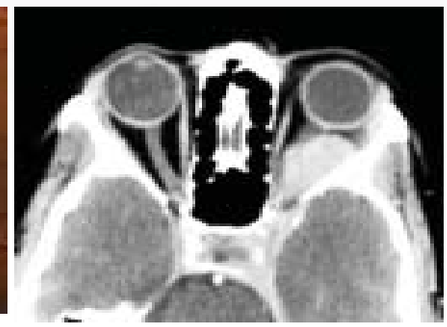


Fig. 5 : Axial CT showed enhancing lesion of left orbit with extension along the superior orbital fissure into the left cavernous sinus

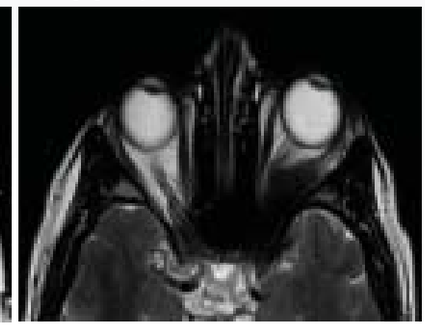


Fig 6. : Axial T2-weighted image through the orbits showing a hypointense, diffusely infiltrative mass lesion in the left orbit.

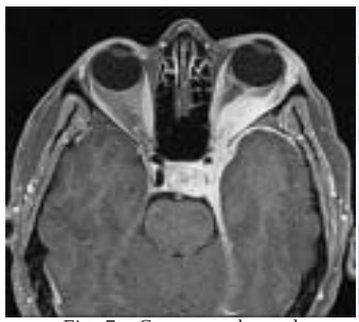


Fig. 7 : Contrast-enhanced T1-weighted images with fat suppression shows an enhancing mass involving the left lateral rectus, superior rectus and the lacrimal

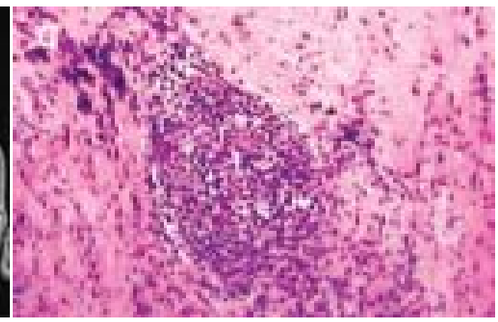


Fig. 8 : Biopsy of fibroadipose tissue in the orbit revealed a mixture of lymphocytes, plasma cells and eosinophils gland, causing proptosis of the globe.

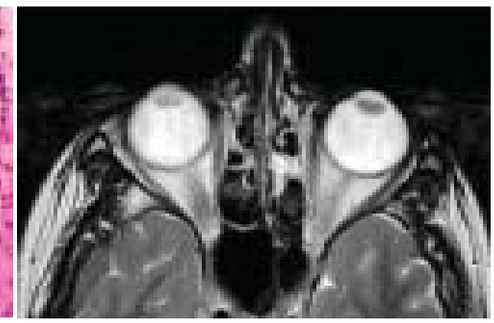


Fig.9 : Follow-up axial T2-weighted MRI showing no signs of the lesion

extended into the optic canal, causing compression and medial displacement of left optic nerve. There was involvement of the left cavernous sinus [Fig. 6, Fig. 7].

Biopsy :

Biopsy was performed via a lateral orbitotomy approach. The histological features were consistent with idiopathic chronic orbital inflammation and fibrosis of adipose tissue and skeletal muscle [Fig. 8].

All the laboratory culture reports were negative. ESR, CRP, and autoimmune screening were normal. Inflammatory workup, including ANA, C-ANCA and RF, were all negative. The chest X-ray was also normal.

The patient was started on 60 mg oral prednisone per day for a week. The dose of oral steroids was then slowly tapered over 4 weeks.

The patient showed improvement in vision, extraocular movements and proptosis on follow-up visits.

Patient's latest visual acuity was 6/9 (OS), with no recurrence to date.

Diagnosis :

A diagnosis of non-specific orbital inflammation was made.

On correlating imaging features, histopathological findings, laboratory workup and response to steroid therapy, the diagnosis was finalized.

Discussion :

Non-specific orbital inflammation (NSOI) is the commonest cause of non-infectious orbital disease. The non-specific presentations of orbital inflammation mimicking other inflammatory processes such as orbital cellulitis and orbital tumors often makes the diagnosis of orbital pseudotumor a challenge to Ophthalmologist. Patients typically present with rapid onset usually unilateral painful proptosis (~90% of cases). The definitive diagnosis without histopathology specimen can be difficult, but a biopsy may not always be amenable. The risk of injuring other orbital structures in the relatively small confinements of intraconal cavity is high. Hence, other diagnostic modalities such as MRI and CT become invaluable tools in aiding the diagnosis.

Conclusion :

Non-specific orbital inflammation can have extraorbital extension resulting in widespread clinical symptoms. In the setting of an orbital mass with intracranial extension, the possibility of pseudotumor should be considered by the reading radiologist. This would not only help the treating physician in the choice of appropriate therapy but also eliminate the need for unwarranted biopsy or surgery.

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