Acute Bilateral Proptosis - A Case Report

Abstract

Orbit truly called the Pandora’s box can reveal many surprises. We present a case of a 45-year-old man who presented with 2 weeks history of protrusion of both eyes after yoga treatment for sinusitis. On examination he had eccentric proptosis, with movement restriction in all gaze and positive FDT. His left eye pupil was sluggish. He had enlarged cervical lymph nodes on left side. Imaging showed features suggestive of fungal sinusitis. As optic nerve head showed compressive changes the patient underwent orbital decompression. The tissue sample was sent for study, which lead to the diagnosis of primary Non-Hodgkin’s Lymphoma.

Introduction

Proptosis could be a silent initial manifestation of serious underlying diseases. Imaging studies alone may not lead to a diagnosis and misleading history can further confuse the picture. Clinical examination coupled with histopathology and an awareness of existence of rare entities will help reach at a diagnosis.

Case Report

A 45 year old man was referred to Ophthalmology OP, Medical College, Kozhikode for evaluation of bilateral acute proptosis. His complaints started with nasal block & nasal discharge of 2 weeks duration and 2-3 episodes of epistaxis & protrusion of both eyes L>R of 3 days duration. He has been on systemic antibiotics and decongestants for his complaints. He was attributing his complaints to some sort of yoga like “Sheershasna” & “Nasyam”. There was no h/o trauma, fever, headache or blurring of vision. He was a chronic alcoholic.

General examination revealed clubbing, bilateral pitting pedal oedema & multiple cervical lymph node enlargement on left side of the neck. Ocular examination showed bilateral proptosis with eccentricity on the left side. There was periorbital fullness, conjunctival congestion and chemosis left > right. Extra ocular movements were restricted, left eye showed positive FDT in all directions. The pupil was sluggish on the left side. Visual acuity, colour vision and visual fields were unaffected. The intraocular pressure was 21 mm of Hg in the left eye and 19 mm of Hg in the right eye. Fundus of the left eye showed hyperemic disc, blurred disc margin, superficial haemorrhage at the inferonasal part of the disc and choroidal folds inferonasal to the disc.

Blood investigations showed altered liver function test, which was attributed to alcoholic cirrhosis. USG abdomen also showed liver cirrhosis with moderate ascites. MRI brain, with orbit and paranasal sinuses revealed lesions with altered signal intensity, without any contrast enhancement, involving all the paranasal sinuses and nasal cavity with intra orbital extension left > right – suggesting a fungal mass with hemorrhage.
Bilateral orbital decompression was done, along with left uncinectomy, middle meatus antrostomy, sphenoidectomy and anterior&posterior ethmoidectomy. Surgery revealed distorted anatomical landmarks with erosion of posterior wall of left maxillary sinus, bleeding polypoidal mass, blackish turbinate mucosa, irregular mass on the septal mucosa. After the surgery proptosis reduced, patient was better and discharged after a course of systemic antibiotics and metronidazole.

To our surprise histopathological report came as chronic inflammatory lesion with necrosis and was advised immunohistochemistry (IHC) to rule out plasmacytoma. But the immunohistochemistry showed positive tumour marker (CD45), negative for B-cells (CD20). Cytokeratin was also negative so as to exclude squamous cell carcinoma. Thus the case was diagnosed as T-cell lymphoma.

The patient was readmitted with features of hepatic failure and septicaemia and succumbed to the disease within 3-weeks.

**Discussion**

Primary Non-Hodgkin’s lymphomas (NHL) of the nose and paranasal sinuses (PNS) are rare, accounting for 0.17-2% of all cases of NHL and 5.8% of all malignant neoplasia of the sinonasal region in adults. The most common paraorbital tumour invading the orbit is squamous cell carcinoma of the PNS constituting 60% of the cases. Out of the lymphomas, more than 50% of ocular adnexal and orbital lymphomas are of Mucosa Associated Lymphoid Tissue (MALT), whereas ocular lymphomas are predominantly Diffuse Large B cell Lymphoma (DLBL). DLBL is a high grade tumour that commonly presents with systemic involvement. B-cell lymphoma of ocular origin are more likely to extend to the orbit than T-cell lymphomas.

T-cell lymphoma in the orbit can present as painful ophthalmoplegia and take a rapid clinical course. The disease should be regarded as one of the differential diagnosis for painful ophthalmoplegia refractory to corticosteroid therapy.

In Asians, T-cell lymphomas are common in nasal cavity, unlike in the western population where B-cell variety predominates. Patients with MALT lymphoma have a more favourable outcome than those with other types of lymphomas.

Common symptoms of orbital and PNS lymphomas are nasal block, nasal discharge, epistaxis, proptosis, diplopia, chemosis and visual defect depending on the optic nerve compression.

Local control of the disease can be obtained by radiotherapy when localized to ocular region. Chemotherapy along with radiation is the treatment in patients with systemic spread.

**Conclusion**

In adults, common causes of acute proptosis are orbital cellulitis, pseudotumour, cavernous sinus thrombosis, CCF, retro-orbital hemorrhage. Here we present, a case of T-cell lymphoma of the nose and PNS which presented as
acute bilateral proptosis. A detailed history, imaging and pathological evaluation is essential to avoid diagnostic dilemmas. So this rare possibility should also be considered while dealing with a case of acute proptosis in adults.

Clinical alertness combined with radiological imaging and pathological tissue diagnosis is essential in early diagnosis and management in all cases of acute proptosis in adults.

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