10.5005/jp-journals-10003-1154

REVIEW ARTICLE

Management Dilemma in a Challenging Case of Inflammatory Pseudotumor of an Orbitomaxillary Mass: A Review of Literature

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ABSTRACT

Objective: To present management dilemma in a challenging case of a benign orbitomaxillary pseudotumor and its importance of inclusion in the differential diagnosis of orbital disorders.

Setting: Private multispecialty hospital in Kolkata, West Bengal.

Patient: A 36-year-old female patient with 1 year history of orbitomaxillary mass, deviation and protrusion of left eyeball, diplopia and left sided headache.

Results: Resection of entire orbitomaxillary mass performed after assessing history, clinical and radiologic presentation of the case. Biopsy report came as mixed inflammatory lesion. Appropriate surgical intervention resulted in a dramatic improvement in patient's clinical condition. At present, patient is disease-free and asymptomatic.

Conclusion: Complete surgical resection is the treatment of choice for inflammatory orbitomaxillary pseudotumors followed by corticosteroids in case of incomplete resection. Radiation therapy is indicated only in case when corticosteroids or surgical intervention is unsuccessful or contraindicated.

Keywords: Diplopia, Inflammatory pseudotumor, Orbitomaxillary mass, Corticosteroids.

How to cite this article: Shah PD, Mukherji S. Management Dilemma in a Challenging Case of Inflammatory Pseudotumor of an Orbitomaxillary Mass: A Review of Literature. Int J Otorhinolaryngol Clin 2014;6(2):61-64.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Inflammatory pseudotumor (IPT) is a benign lesion with no identifiable local or systemic cause. First described in 1905 by Birch-Hirschfield.[1,2] The term 'pseudotumor' was used because these lesions mimic invasive malignant tumors both clinically and radiologically. It is also known as idiopathic orbital inflammatory syndrome. It is a nonspecific, non-neoplastic inflammatory process of the orbit.[1-4] It accounts for only 4.7 to 6.6% of all orbital diseases. Inflammatory pseudotumor most commonly involves the lung and orbit but has also been reported to occur at sites that make biopsy or excision difficult or potentially disfiguring.[5] An orbitomaxillary mass may primarily be an orbital lesion extending into the maxillary sinus or a primary maxillary sinus lesion extending into the adjacent orbit.[6] The behavior of IPT can be quite unpredictable. Some resolve spontaneously and others respond to corticosteroids. In some cases, tapering steroid doses cause lesions to recur. For lesions not responding to steroids, excision of lesion with or without radiotherapy may be required.[7]

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CASE REPORT

A 36-year-old female patient reported to Department of Oral and Maxillofacial Surgery, Fortis Hospital, Kolkata with complaining recurrent swelling and pain over left eye since one and half years and also complained of upwards deviation and protrusion of left eye ball which was progressively increasing, left sided headache and double vision (Fig. 1). After obtaining an informed and written consent regarding inclusion in study, local and general physical examination carried out. On examination patient had restricted left eyeball movement in horizontal gaze and diplopia was noted. Extraoral swelling and tenderness over left cheek was evident but patient denied excessive lacrimation, blurring of vision, eye discharge or numbness. Brisk pupillary reflex to light was similar in both the eyes.

Patient sought medical attention at a local hospital and she was referred to various specialties like ENT, ophthalmology, neurosurgery and general medicine. An orbital CT scan and MRI were taken on advice of ophthalmologist and provisionally diagnosed as fibrous dysplasia of orbital bone. Patient was advised for incisional biopsy of maxillary sinus mass through endoscope and orbital mass through anterior orbitotomy approach under general anesthesia. Patient was given oral prednisolone for 3 months; initial dose was 60 mg/day for 1 month following every month dose was tapered to 40 mg/day and 20 mg/day. Partial improvement was noted on taking oral steroids. There was no significant medical history in the past.



Fig. 1: Proptosis and upward shift of left globe with diplopia

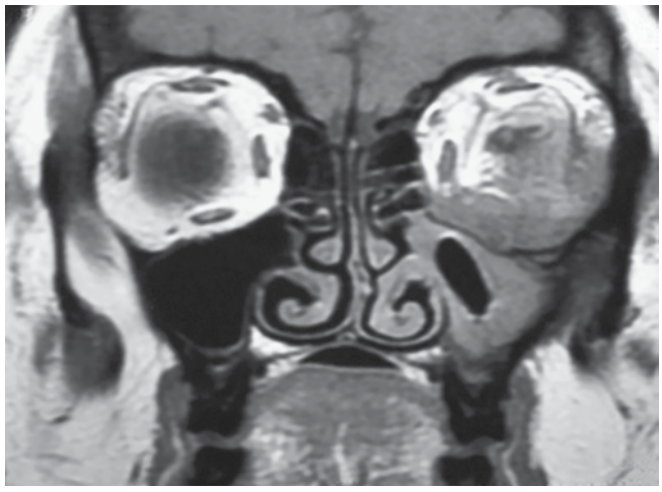


Fig. 2: MRI showing tumor invading left maxillary sinus and extending to left orbit

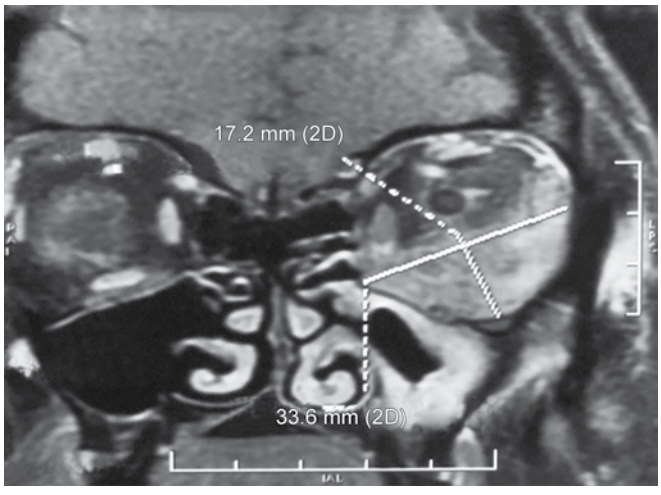


Fig. 3: MRI depicting the size of lesion



Fig. 4: Orbital extenuation of tumor grabbed

On general physical examination, patient was found to be hypertensive. All routine blood investigations, serology tests were performed. Patient's fasting blood sugar was 250 mg/dl which was significantly high. MRI showed expansile, mildly enhancing soft tissue mass with peripheral foci of calcifications occupying the maxillary sinus and extending up to postero-inferior aspect of the left orbital cavity (Figs 2 and 3).

As patient had high blood sugar, we decided to stop oral prednisolone and to go for complete resection of orbito-maxillary mass. Patient was drapped and prepared under standard surgical protocol. Lateral canthotomy and cantholysis were performed; incision was deepened through skin, muscle, lateral canthus and conjuctiva. Transconjunctival incision performed till medial canthus. Periosteum identified and incised. Subperiosteal dissection performed postero-inferiorly. Periorbital periosteum was gently separated from extraoccular muscles and delivered out (Figs 4 and 5). Access to maxillary sinus was gained through Caldwell Luc approach. 2.5 × 1 cm diameter bony window was created and mass was delivered out. Remaining bits of maxillary sinus mass was identified using endoscope and brushed out (Fig. 6). Specimen was sent for histopathological examination. Buccal bony window was replaced and fixed using 1 × 3D plate and 4 × 6 mm length titanium screws. Intraoral closure was performed using vicryl 3-0. Lateral palpebral conjuctiva was closed with vicryl 6-0. Lateral canthotomy incision was closed using Prolene 6-0.

Postoperatively significant improvement was observed by the patient, left and right eyeball were nearly at same level. There was no sign of diplopia. Duration of hospital stay was uneventful and patient was stable and discharged on 5th postoperative day with advice to take salt restricted diabetic diet.

Microscopic examination revealed mixed inflammatory lesion. Sections of specimen showed hyalinized fibro-collagenous tissue, blood clot, lamellar bone and respiratory mucosal fragments, lamina propria of which showed



Fig. 5: Orbital tumor completely resected via transconjunctival approach

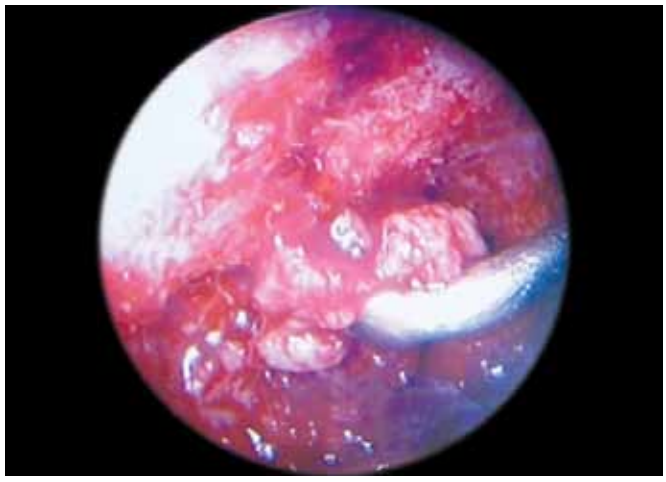


Fig. 6: Maxillary sinus mass identified using endoscope and brushed out



Fig. 7: Before and after

multifocal dense, sheet-like mixed inflammatory cell infiltrates comprising of neutrophils and lymphoplasmacytic cells. Neutrophilic exocytosis into ciliated columnar mucosal glands was noted. Accompanying fibrin exhibited prominent neutrophilic inflammatory cell infiltration at places. Malignancy was not seen.

When patient was seen 3 months postoperatively, there was absence of periorbital swelling, eyeball rotation, diplopia and restricted eyeball movement. Left and right eyeball were nearly at the same level (Fig. 7).

DISCUSSION

Orbital mass lesions may arise primarily within the orbit, extend from contiguous structures, or be metastatic from a distant primary malignancy. Primary orbital masses include congenital, vascular, neural and mesenchymal tumors. Other types of lesions masquerading as true neoplasms include lymphoproliferative disorders, autoimmune diseases and infectious processes. IPT is an idiopathic inflammatory lesion although various stimuli may cause it to develop such as unrecognized organisms, minor trauma, smoking, and chronic irritation by cocaine abuse.[8] The exact etiology of these lesions is not clear. It has been postulated that they might be the result of a postinflammatory repair process, a metabolic disturbance or an antigen-antibody interaction with an agent that was no longer identifiable in aspiration or biopsy material.[6] Some authors relate IPT to production of mediators of inflammation which stimulate proliferation of fibroblasts, extravasation of neutrophils and activation of procoagulant activity of the vascular endothelium. The clinical findings in a patient with an inflammatory pseudotumor are variable depending on the growth rate of the lesion and the specific structures that have been affected. Extraorbital inflammatory pseudotumor of head and neck can occur in the nasal cavity, nasopharynx, maxillary sinus, larynx and trachea. Perineural spread along maxillary, mandibular and hypoglossal nerves had been described. Sinonasal inflammatory psuedotumors do not affect a particular age

group and cause no systemic symptoms. However, they have a more aggressive appearance than those of the orbit with bony changes such as erosion, remodeling and sclerosis usually seen on radiographic studies.[9] On imaging studies, patients may show stigmata of orbital inflammation, including fluid in the Tenon's space, molting of the optic nerve contour and thickening of the extraocular muscles.[10] On CT-scan and MRI studies, pseudotumors may present with diffuse orbital mass, uveoscleral thickening, contrast enhancement of Tenon's potential space, proptosis and optic nerve and extraocular muscle enlargement.[10,11] In some patients, laboratory findings are normal; in others, there might be an elevated erythrocyte sedimentation rate and C-reactive protein level and sometimes a high white blood cell count.[12] However, none of the published reports on inflammatory pseudotumor have mentioned any presence of positive tumor markers.[6]

CONCLUSION

Understanding of the clinical features of patients with orbital pseudotumor, differentiating it from other orbital processes by use of imaging techniques and timely implementation of available treatment strategies may help prevent visual loss and associated morbidity from this condition. If possible complete surgical resection is the treatment of choice for orbitomaxillary inflammatory pseudotumors, followed by corticosteroids in cases of incomplete excision. Response to steroids is often unpredictable. Radiation therapy is indicated only in case when corticosteroids or surgical intervention is unsuccessful or contraindicated.[13]

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