IDIOPATHIC ORBITAL INFLAMMATION (PSEUDOTUMOR)

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INTRODUCTION

Orbital pseudotumor is uncommon but not rare. In 1930 Birch-Hirschfeld first used the term pseudotumor to describe orbital inflammation that was associated with proptosis and appeared to be caused by a neoplasm. He described different types of "pseudotumor" [1].

Orbital pseudotumor is now considered as a nonspecific, idiopathic, benign inflammatory process consisting of lymphocytes, plasma cells, fibroblasts, macrophages, occasional neutrophils, eosinophils, and epithelial cells; rare granuloma formation and lymphoid follicles; and collagen deposition, localized to the orbit. The clinical course of the disorder may be acute, subacute, or chronic. There is no sex predilection.

We are reporting a case to emphasize that it is important in differential diagnosis of proptosis.

CASE REPORT

A 43 years old man, tailor by profession and working in the Gulf, presented with painful swelling of right eye, decreased vision and divergence of right eye two weeks back. He had similar episode 10 months ago for which he remained for 2 months on daily dose of 60 mg of oral prednisolone and developed mild Cushingoid features (weight increased from 95 to 103 kg). He was a diagnosed hypertensive and his BP is 160/110 mm of Hg with cap Amlodipine Besylate (Norvasc) 5 mg daily. He is a smoker, smoking 20 cigarettes in 1-2 days. There was no history of cough or nasal problems. On examination of right eye there was lid swelling, conjunctival congestion and divergence of 30 degrees. With Hertel exophthalmometer, proptosis was 5 mm. There was severe restriction of elevation and adduction of right eye. Vision was 6/24, not improving with pin hole and there was no refractive error. Fundus examination revealed swelling of optic disc with haemorrhages and cotton wool spots. Intraocular pressure was 24 mm of Hg. On palpation the orbital pressure was raised, proptosis was non reducible and there was no thrill or bruit. Left eye was normal with unaided visual acuity of 6/6 and 15 mm of Hg IOP.

Ultrasound B scan of right orbit revealed hypo-echoic (solid), intraconal mass medial to the eyeball. Doppler scan showed minimal vascularity. CT Scan (axial and coronal views with 3 mm sections) (Fig. 1) confirmed the findings of ultrasound. Medial and Lateral recti were displaced away from the mass. Lateral rectus was slightly thickened. There was no calcification, no hyperostosis, optic canal was not widened. In ethmoid sinuses, a few cells were hazy. Right maxillary sinus was small and hazy. Contrast uptake by the mass was minimal. X-ray Chest (PA view) did not reveal any abnormality excluding pulmonary tuberculosis and sarcoidosis.

He was started on 30 mg of oral Prednisolone and given 40 mg posterior subtenon injection of Methyl Prednisolone (Depo-medrol) which resulted in marked improvement in conjunctival congestion, ocular motility and vision (improving to 6/6). Incisional biopsy (4cc of mass removal) was done after disinserting anterior attachment of medial rectus. It corrected the divergence also (Fig.2). Adipose tissue, having a few fibrous septa, was found on histopathological examination. The patient was maintained on low dose oral steroids (10 mg of Prednisolone daily) which kept him asymptomatic for one month after which he did not report back.

DISCUSSION

Pseudotumor has been observed in children as young as 9 years [1] and even 2 years [2]. It was observed in one study that mean age of presentation was 43.75 years, 73.5% had unilateral disease, the clinical

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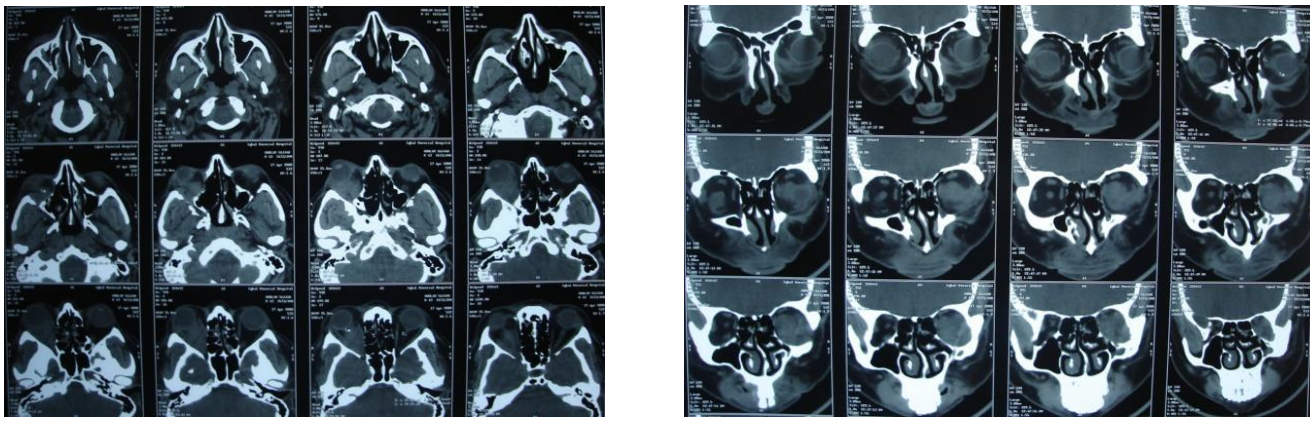

Fig. 1: CT Scan (left) axial and (right) coronal views. Isodense, SOL was seen in retro bulbar and below the optic nerve. Normal Med rectus and slightly thickened Lateral rectus were displaced away from the mass.

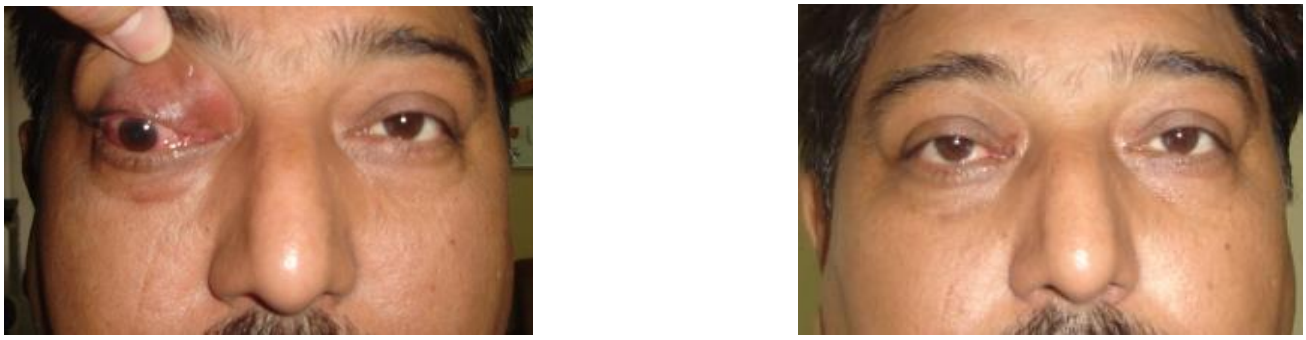

Fig. 2: Left: Right eye shows lid swelling and erythema, conjunctival congestion, inferior dystopia and divergence of 30 degrees, Right: After treatment. Lid and eye ball position - normal

features were proptosis (79.6%), ocular motor deficit (61.2%), pain (51%), lid swelling or lid mass (44.9%), ptosis (24.5%), chemosis (18.4%) and improvement occurred in 81.6% patients with corticosteroids with mean recovery time of 10.3 days for visual loss. Ocular motility recovered in 80% patients, an average of 17.8 days after initiation of therapy [3]. There may be extraorbital extension of inflammation [4]. 15% of orbital masses may prove to be because of idiopathic inflammation [5].

The most important investigations to be performed for evaluating idiopathic orbital inflammation are orbital B – Scan ultrasonography and CT Scan. PCR may be required to identify mycobacterium DNA [6]. One of the important diagnostic aids in the evaluation of idiopathic orbital inflammation is the use of a trial of high-dose corticosteroids. This disease is almost always responsive to systemic corticosteroids. There is a danger of developing corticosteroid dependence [7]. Intraorbital triamcinolone has been found effective [8].

Chronic orbital lesions not responding completely to corticosteroids should undergo biopsy to establish a tissue diagnosis of nonspecific inflammation and to exclude specific inflammations. The majority of biopsies may be performed via an anterior orbitotomy under general or local anesthesia.

Surgical excision to debulk the tumor mass is required to control aggressive lesions followed by systemic steroids in tapering dose and low maintenance dose. Refractory cases may be better managed by other modalities like radiotherapy or chemotherapy.

Our patient had visual deterioration, eye inflammation, proptosis, dystopia and weight gain which drastically affected his quality of life. History, clinical features and ultrasound/ CT scan were in favour of orbital inflammation. History of good response to steroids in the past, made neoplastic causes unlikely. Presence of orbital inflammation and sinusitis on CT scan raised the doubt of Wegner's granulomatosis. However facts making this diagnosis unlikely, were unilateral involvement and that there

were neither upper respiratory symptoms like nasal obstruction, serosanguineous discharge from the nose or lesions of the mucosa nor symptoms of lung involvement which include cough, dyspnea, chest pain or hemoptysis. There were no skin lesions.

Our patient was initially managed with moderate dose of oral corticosteroids which resulted in significant improvement. As patient was upset with oral steroids, subtenon injection was given which has been used rather infrequently as is obvious from the literature. Later to further reduce the proptosis as well as to rule out infectious entities and various vasculitides, incisional biopsy / mass debulking was done. It corrected the eyeball position completely. Histopathology report was negative for infectious entities. Absence of inflammatory reaction and various vasculitides may be because of non representative area or because of course of steroids. It is known that even in patients with a well-documented diagnosis of Wegener's granulomatosis, orbital biopsy may not demonstrate the classic features of vasculitis, granulomatous inflammation, and tissue necrosis in as many as 50% of cases [9]. Low dose of maintenance corticosteroids were required to keep him asymptomatic. Longer follow up of patient was desired but unfortunately he went back and could not be contacted again.

Orbital pseudotumor is an important cause of proptosis which may be associated with significant morbidity. Specific causes of inflammation should be excluded. Treatment with corticosteroids results in prompt and marked improvement. Posterior subtenon route is also effective.

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