

## Case Report

# A Case Study on Idiopathic Orbital Pseudotumor: Surgery and Steroid Treatment

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### Abstract

*This is a case study of five isolated orbital nerve inflammatory pseudotumor cases presenting with protrusion and visual acuity of the right eye. Optic disk edema was observed by ophthalmoscopy. Plain and contrast-enhanced magnetic resonance imaging (MRI) were used to examine the orbital fat and enlargement of the right optic nerve sheath along with orbital magnetic resonance imaging diagnostic test. The visual acuity (VA) of all the admitted patients was 1.5/20. All the patients reported attacks of retrobulbar pain and severe headache at fairly regular intervals. The patients on clinical examination were found to have protrusion, reduced visual acuity of the right eye with an ipsilateral afferent pupillary defect. There was no motility of the right eye and the exophthalmos of the right part was 27 mm. On examination, patients were diagnosed to have idiopathic orbital inflammation and received steroid therapy for 6 months. Thereafter, patients received surgical treatment to remove the superior wall of the orbit followed by cortisone therapy for a further 6 months. The follow-up examination of the patients for one year revealed a gradual improvement in the vision of all the patients. The VA of the right eye for all the patients was 6/20, and the exophthalmos was now around 19 mm. Thus, a dramatic response to surgery and steroid treatment was observed in all the patients.*

**Keywords:** Orbital fat, Visual acuity, Optic disk edema, Optic nerve, Cortisone therapy

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## INTRODUCTION

Idiopathic orbital pseudotumor (IOP) is an inflammation arising due to nongranulomatous orbital mass [1] and accounts for 4.7-6.3 % of all orbital disorders [2,3]. It is usually observed after 45 years of age [4]. After thyroid ophthalmopathy and lymphoproliferative disorders, IOP is the third commonly observed orbital disease. The histopathological examination of IOP revealed the presence of fibro inflammatory infiltrate and lesions with atypical histopathologic patterns [2]. It is found in the orbit where main target is the lacrimal gland [4]. IOP is mostly unilateral [1,4]

and the symptoms include diplopia, conjunctival chemosis, visual disability and restriction of extraocular muscle function [4]. IOP comprises different categories of orbital inflammatory diseases involving various levels of orbital contents. The optic nerve may be affected as a part of the posterior segment process [5]. Isolated optic nerve is very rarely involved and has been shown to be involved in one patient out of 16 cases of orbital pseudotumor by Peyster *et al* [6].

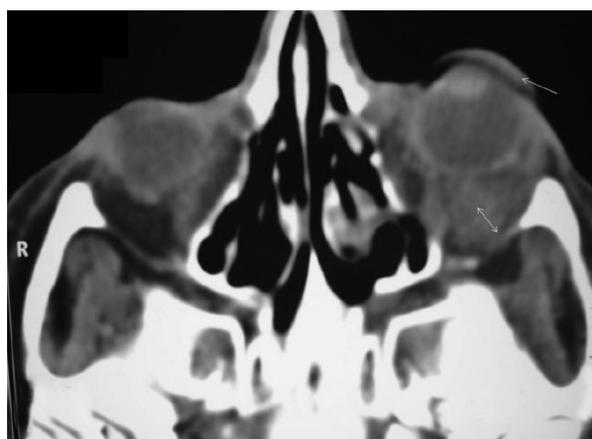
The inflammatory process may affect different orbital structures like the globe, extraocular

muscles, lachrymal glands and the optic nerve [7]. Deposition of fibro collagen over the affected nerve/ sheath complex leads to diffuse enlargement of the optic nerve [8-10]. Idiopathic orbit inflammatory pseudotumor is usually associated with other diseases, including, Tolosa Hunt syndrome, pituitary histiocytosis, idiopathic meningitis, carotido-cavernous fistula, Wegener granulomatosis and Erdheim–Chester disease [11,12]. The study was performed to investigate the efficacy of surgical extraction of the superior wall of orbit followed by cortisone therapy for 6 months in the improvement of vision of patients with protrusion and visual acuity.

## EXPERIMENTAL

### Patient

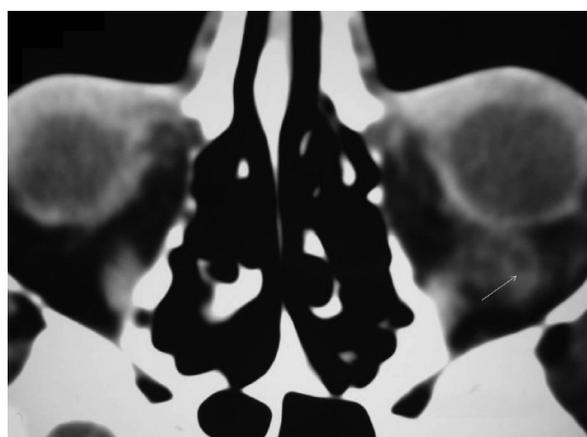
Five patients, 3 men and 2 women (average age 53 year) with more than one-year history of progressive protrusion and reduced vision of the right eye were admitted to our hospital (Figure 1). The visual acuity (VA) of all the patients was 1.5/20.



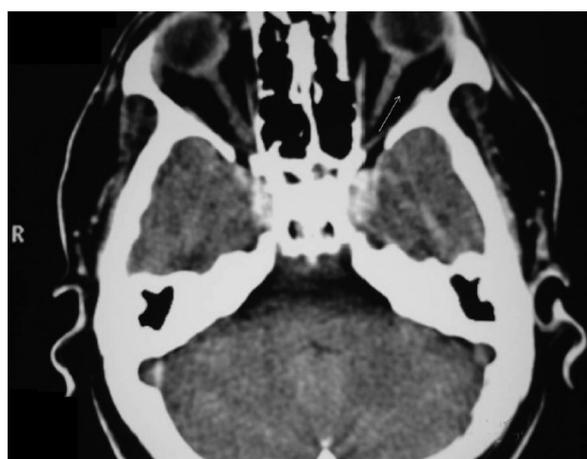
**Figure 1:** Pre-treatment-Contrast enhanced CT scan of orbit axial view showing anterior displacement of the right globe (arrow) and non-enhancing soft tissue mass in the right orbit (double arrow)

Careful examination of the patients revealed absence of any trauma to the orbit. All the patients reported attacks of retrobulbar pain and severe headache after regular intervals. The patients on clinical examination were found to have protrusion of the right eye, reduced visual acuity of the right eye with an ipsilateral afferent pupillary defect. There was no motility of the right eye and the exophthalmos of the right part measured 27 mm. We also observed a central scotoma and poor color perception in all the patients. Optic disk edema was observed on examination by Ophthalmoscopy. Serum levels

of antinuclear antibody were normal. However, diffuse extra- and intraconal mass infiltrating the orbital fat and enlargement of the right optic nerve sheath of the orbit in all the patients was clearly seen. The cavernous sinuses and the brain were normal. On examination patients were diagnosed to have idiopathic orbital inflammation and received steroid therapy for six months. Following chemotherapeutic treatment patients underwent surgery for extraction of superior wall of the orbit. After surgery cortisone therapy was continued for six months more. Follow-up examination of all the patients for one year revealed improvement in the vision. The VA (Functional Vision Analyzer from Stereo Optical Company, Inc. Chicago, United States) of the right eye for all the patients was 5/20, and the exophthalmos was measured to be around 19 mm (Figures 2, 3).



**Figure 2:** After 6 months of starting steroid therapy contrast enhanced CT Scan of right orbit axial view showing partial resolution of the soft tissue mass



**Figure 3:** After 1 year of treatment- contrast enhanced CT scan of orbit- axial view showing complete resolution of proptosis and soft tissue mass

Dense fibrous proliferations, with collagen deposition, infiltrating the muscles were observed during histopathological examination of the biopsy. Numerous inflammatory cells were observed, consisting mainly of lymphocytes, diffusely disposed or arranged in follicles with germinal centers. Plasma cells, macrophages and scattered neutrophils were also observed. A granulomatous inflammatory reaction was present in some areas, with epithelioid cells and giant multinucleated cells.

## DISCUSSION

Idiopathic orbital pseudotumor is associated with symptoms similar to many other intraorbital tumors [8] making the diagnosis difficult. Image findings are very useful for the characterization of the inflammatory process level. In IOP the commonly observed features include retro-orbital fat infiltration, proptosis, extraocular muscle enlargement and enhancement, orbital apex inflammation and optic nerve thickening [13]. Edematous enlargement of the optic nerve in the perineuritic form of the disease is shown by computer tomography (CT) and magnetic resonance imaging (MRI). The optic nerve/sheath complex enlargement is caused by the neoplastic and non-neoplastic conditions [10]. The neoplastic causes include optic gliomas, meningiomas, neurofibromas, hemangioblastomas, metastases, leukemias and lymphomas. On the other hand, non-neoplastic causes include Optic neuritis, pseudotumours, granulomatous optic neuropathies (most commonly sarcoidosis), dysthyroid orbitopathies, traumatic hematomas and optic nerve drusen.

Optic nerve drusen is associated with pseudopapilloedema and deposition of calcium between the optic nerve and globe. In the present study, tumor cases were excluded because of absence of the optic canal enlargement [14], hyperostosis and calcification [15-17]. However the symptoms of orbital pain and headache were considered to be of inflammatory etiology [10-14]. Finally, the results from surgery of the orbit and the response to a therapeutic trial of steroids clinched the diagnosis in favor of inflammatory pseudotumor [18-21].

## CONCLUSION

Surgical extraction of the superior wall of orbit followed by cortisone therapy for 6 months improved the vision of patients with protrusion and visual acuity of the right eye. Thus, the strategy can be used for treatment of the patients with orbital protrusion and visual acuity.

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