

Orbital compartment syndrome in idiopathic orbital inflammatory disease: A case report

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SUMMARY

Orbital compartment syndrome (OCS) is a visual threatening ocular emergency. We report a 50-year-old male with acute presentation of OCS, a rare manifestation of idiopathic orbital inflammatory disease. At presentation, high intraocular pressure was reduced by prompt lateral canthotomy and cantholysis. The disease responded to systemic steroids and treatment resulted in good visual outcome. Detail evaluation and early detection and treatment are mandatory to prevent permanent vision loss.

KEY WORDS:

Orbital compartment syndrome, Idiopathic Orbital Inflammatory Disease

INTRODUCTION

Idiopathic orbital inflammatory disease (IOID) is a rare inflammatory disorder of the orbit. Nevertheless it is reported to be the third most frequent orbital disorder following thyroid orbitopathy and lymphoproliferative disease.¹ Orbital compartment syndrome (OCS) is uncommon and occurs due to an increased pressure within the confined orbital space from either a mass or swelling of the orbital contents. It may potentially lead to blindness due to compression of the optic nerve that lies within the orbital cavity, but vision can still be salvaged with prompt treatment. We report a rare case of an acute OCS secondary to IOID.

CASE REPORT

A fifty-year-old healthy male presented with left eye acute proptosis with sudden onset blurring of vision and pain, upon waking up in the morning. There was no history suggestive of thyroid eye disease or orbital cellulitis. Patient was afebrile. Examination of the left eye showed visual acuity of 6/60. The orbit was tight with a non-axial proptosis, chemosis, inflamed conjunctiva and a large exposure keratopathy (Figure 1). There was a positive left relative afferent pupillary defect detected. The left intraocular pressure (IOP) was 50 mmHg however fundus showed no abnormality. The right eye and systemic examinations were unremarkable.

An immediate left lateral canthotomy and cantholysis were performed which instantly reduced the IOP to 20 mmHg. Blood tests which included full blood count, renal profile, liver function test, erythrocyte sedimentation rate were

normal. Computed tomography of orbit showed left irregular outline of the optic nerve and medial rectus muscle thickening with retro-orbital fat stranding; suggestive of inflammatory changes in the retro-orbital region. Magnetic resonance imaging of the orbit confirmed the diagnosis of left IOID. Based on the above, patient was diagnosed to have left acute OCS secondary to IOID. He was treated with intravenous methylprednisolone 1 g daily for 3 days followed by oral prednisolone 1 mg/kg/day. Topical lubricants were given. His visual acuity improved to 6/6 within a week with resolution of proptosis, chemosis and IOP.

DISCUSSION

IOID; also known as orbital pseudotumour, is an uncommon non-infectious, non-neoplastic inflammatory disorder with unknown aetiology. It accounts for approximately 6% to 8% of all orbital inflammatory lesions.² The peak incidence is between the 4th and 5th decades with no gender or ethnic predilection. IOID may involve any part of the orbital soft tissues. It is the most common cause of painful orbital mass in adults. Proptosis followed by restrictive myopathy is the most common presentations, as demonstrated in this patient. The pathogenesis of IOID remains unclear, however it is reported to have an association with Immunoglobulin G4 related disease.³ Mombaerts I et al. found that autoimmune disorder, infection and aberrant wound healing may play important roles in its pathogenesis.⁴

IOID is a diagnosis of exclusion. Infective e.g., orbital cellulitis, and non-infective causes e.g., thyroid eye disease, sarcoidosis, granulomatosis with polyangiitis, malignancy and vascular malformation, of orbital inflammation, need to be carefully ruled out initially. Meticulous clinical evaluation and thorough investigations are therefore mandatory. Computed tomography scan of IOID usually shows ill-defined orbital opacification. The medial rectus muscle is commonly affected as seen in this case.⁵

Corticosteroid is the mainstay of treatment for IOID and provides favourable and quick outcome particularly in acute presentation.¹ However, approximately half of the patients may develop recurrence which may be contributed by rapid tapering of corticosteroid treatment. Radiotherapy is used in conjunction with steroid when steroid treatment alone fails.¹ Immunosuppressants may have a role when there is poor response to steroid and radiotherapy.

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Fig. 1: Left eye showed chemosis, proptosis and exposure keratopathy.



Fig. 2: Left eye resolved proptosis.

OCS is an ocular emergency. Irreversible visual loss may result as early as 30 minutes if left untreated. This condition may occur secondary to retro-orbital haemorrhage, orbital emphysema, orbital cellulitis, orbital abscess or tumour. To the best of our knowledge, to date, there is no published literature of orbital compartment syndrome secondary to IOID yet. In IOID, excessive orbital inflammation leads to an increase in orbital volume within the enclosed orbital space resulting in proptosis, increased IOP, optic nerve damage and eventually OCS. Urgent orbital decompression is the mainstay of treatment in OCS. Lateral canthotomy and cantholysis are usually performed to sufficiently relieve the enclosed orbital space, allowing the orbital volume to expand anteriorly and reducing the compression to the optic nerve.

CONCLUSION

IOID is a heterogeneous group of ocular inflammation with various presentations. It rarely present with OCS. Careful assessment and early detection of OCS are essential to avoid irreversible loss of vision.

REFERENCES

1. Yuen SJ, Rubin PA. Idiopathic orbital inflammation: distribution, clinical features, and treatment outcome. *Arch Ophthalmol* 2003; 121(4): 491-9.
2. Chen YM, Hu FR, Liao SL. Idiopathic sclerosing orbital inflammation - a case series study. *Ophthalmologica* 2010; 224(1): 55-8.
3. Berry-Brincat A, Rose GE. Idiopathic orbital inflammation: a new dimension with the discovery of immunoglobulin G4-related disease. *Curr Opin Ophthalmol* 2012; 23(5): 415-9.
4. Mombaerts I, Goldschmeding R, Schlingemann RO, *et al.* What is orbital pseudotumor? *Surv Ophthalmol* 1996; 41(4): 66-78.
5. Pemberton JD, Fay A. Idiopathic sclerosing orbital inflammation: a review of demographics, clinical presentation, imaging, pathology, treatment, and outcome. *Ophthal Plast Reconstr Surg* 2012; 28(1): 79-83.