

Case Report

Idiopathic Orbital Inflammatory Pseudotumour Mimicking Optic Nerve Sheath Meningioma- A Case Report

Umi Kalthum MN¹, Amin A¹, Syazarina SO², Faridah HA¹

¹Department of Ophthalmology, ²Department of Radiology, Universiti Kebangsaan Malaysia, Jalan Yaacob Latiff, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia.

Abstract

A 42-year-old Malay gentleman presented with progressive painless blurring of vision on his left eye associated with ocular ache, redness and increasing proptosis. Examination revealed presence of relative afferent papillary defect with visual acuity of counting finger, and 6/6 on the right eye. Extraocular muscle movement of the left eye was restricted to ten percent in all directions. Computed tomography (CT) and magnetic resonance imaging (MRI) showed enhancing mass occupying the left orbital apex. Diagnosis of optic nerve sheath meningioma was made, and patient subsequently went for radiotherapy. His symptoms subsided completely, until he presented with similar visual complains, and fullness of the upper lid two years later. A repeated MRI showed enlargement of superior and lateral recti muscles with extension to lacrimal gland region. Biopsy of the lacrimal gland revealed inflammatory cells consistent of inflammatory pseudotumor. High dose systemic steroid was instituted, followed by slow tapering of oral steroid. His symptoms completely resolved, and latest visual acuity was 6/9, with no recurrence, to date.

Keywords: Orbital inflammatory pseudotumour, optic nerve sheath meningioma, magnetic resonance imaging, steroid, radiotherapy

Correspondence:

Umi Kalthum Md Noh, Department of Ophthalmology, Universiti Kebangsaan Malaysia, Jalan Yaacob Latiff, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel no: 03-9145 5981 Fax: 03-9145 6733 Email: umi_fizul@yahoo.com

Date of submission: 10 Nov, 2012

Date of acceptance: 19 Feb, 2013

Introduction

Idiopathic orbital inflammatory syndrome presents with a variety of non-specific clinical symptoms, making diagnosis a challenge in majority of the cases. Conditions such as orbital neoplasm and orbital cellulitis always need to be excluded to eliminate possibilities of life threatening or other vision threatening diseases. Although, biopsy is the most ideal method of confirming the diagnosis, this may not always be amenable as the risks of injuring other orbital structures. Magnetic resonance imaging (MRI) has become the most important diagnostic tool in making the diagnosis (1). However, orbital cellulitis, orbital lymphoid lesion and orbital inflammatory syndromes are not readily differentiated in (MRI) (2). We report a case of idiopathic orbital pseudotumor initially diagnosed as optic nerve sheath meningioma, highlighting the diagnostic challenge in such cases.

Case Report

A 42-year-old Malay gentleman presented with progressive painless blurring of vision on the left eye, associated with periocular ache, diplopia and increasing eye redness. This was his first episode, and he was asymptomatic prior to this presentation. There were no constitutional symptoms reported. Visual acuity at presentation was counting finger, with marked relative afferent papillary defect on the left eye. Extraocular muscle movements were restricted to ten percent on the left eye with diplopia at all gazes. The left optic disc was markedly swollen (Fig.1). Urgent CT and MRI revealed enhancing mass occupying the left orbital apex (Fig. 2). Following discussions with neurosurgery and radiology specialist, a diagnosis of optic nerve sheath meningioma was made. He completed twelve cycles of radiotherapy with a total of 2500 rads delivered. There

was complete resolution of symptoms and visual acuity improved to baseline.

Two years later, he came with similar complains with additional fullness of the upper lid region. A repeat MRI showed enlargement of the medial and lateral recti, enhancing lacrimal gland and periorbital fat streakiness (Fig. 3). Biopsy of lacrimal gland confirmed lymphocytic infiltrates, consistent with orbital pseudotumor. The patient was started on intravenous methylprednisolone 1g/day for three days, followed by oral steroid with slow taper. To date, he is symptom free and visual acuity remains good.

Discussion

Orbital inflammatory pseudotumor is the commonest cause of noninfectious orbital disease (3). The nonspecific presentations of orbital inflammation mimicking other inflammatory processes such as orbital cellulitis and orbital tumors often makes the diagnosis of orbital pseudotumor a challenge to Ophthalmologist. The definitive diagnosis without histopathology specimen can be difficult, but a biopsy may not always be amenable. The risk of injuring other orbital structures in the relatively small confinements of intraconal cavity is high. Hence, other diagnostic modalities such as MRI and CT become invaluable tools in aiding the diagnosis.

Optic nerve sheath meningioma or intraorbital meningioma is a benign, yet an important cause of visual loss secondary to optic nerve sheath compression. Although most commonly, affecting the females in 40s or 50s years of age, it may still occur in different subgroup of patients. In cases of orbital meningioma which compromise optic nerve and hence, vision, the treatment option is limited to radiotherapy as surgical resection of tumour may damage to optic nerve itself.

MRI has become a valuable tool and the procedure of choice in the diagnosis of both the orbital pseudotumor and optic nerve sheath meningioma (4). Radiological findings in orbital pseudotumor involve inflammatory changes of intraorbital structures including the optic nerve, periorbital fat, extraocular muscles and lacrimal glands. Optic nerve sheath meningioma commonly exhibit calcification, which is best visualized in CT scan. Typically, meningioma lesion is isointense on T1 weighted images and hyperintense on T2 weighted images, compared to optic nerve on the MRI. However, this case demonstrated that it may not always be true. The differentiation of meningioma from orbital pseudotumor which relies heavily on MRI findings is



Figure 1: Photograph of fundus shows marked left optic disc swelling with minimal optic disc haemorrhage.

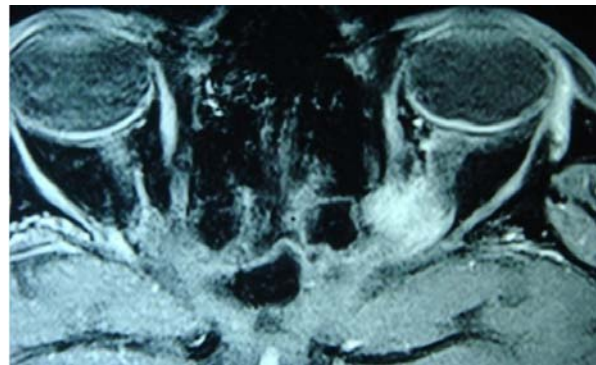


Figure 2: MRI-T1 contrast image on axial view shows enhancing mass at the left orbital apex.

necessarily difficult if structures other than optic nerve are not involved. Subtle areas of enhancing lesion or inflammation may not be appreciated on conventional MRI due to the high signal intensity of surrounding orbital fat. Selective fat saturation and gadolinium contrast should be used to diminish the fat signal and maximize orbital anatomy (5). Diffusion-weighted image has been shown to be useful in differentiating orbital pseudotumor from orbital cellulitis and lymphoid tumour, hence more rapid management is needed (6). Important differential diagnoses to exclude are orbital cellulitis, orbital neoplasms and thyroid orbitopathy.

Therapy is aimed to improve vision, reduce inflammatory symptoms and preserve globe functions. Orbital inflammatory pseudotumor shows dramatic and excellent response to steroid, (7) but some cases may recur or become steroid dependent. High dose of initial steroid followed by slow taper is usually associated with gastrointestinal side effects and reduction in bone density, and evaluations for adverse

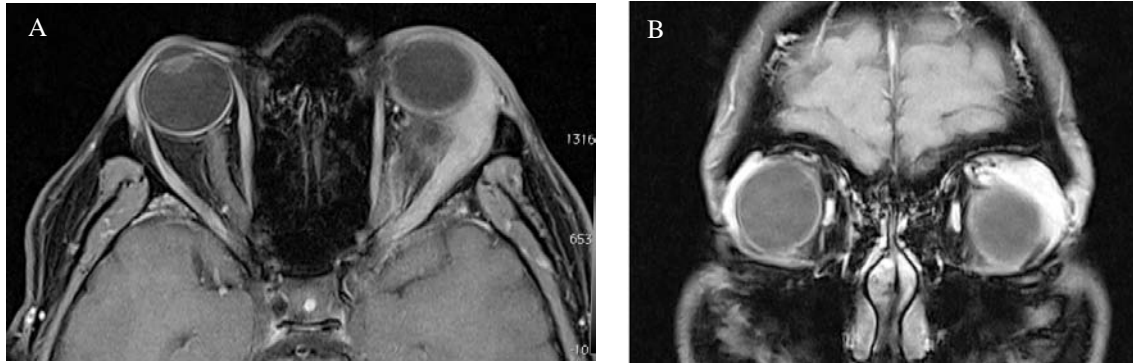


Figure 3: Axial (A) view of MRI- T1 weighted fat- suppressed post- contrast image shows enlargement of medial, lateral recti and periorbital soft tissue. Coronal (B) view of T1- weighted fat- suppressed post- contrast image shows similar enhancement of lacrimal gland and superior rectus- levator palpebrae superioris complex. Enhancement of retro- bulbar.

effects of steroids is mandatory. Radiotherapy is not the initial choice of treatment for orbital inflammatory pseudotumor (8). Although, good response to radiotherapy has been demonstrated on myositis cases, recurrences have been reported (9). Adequate response to radiotherapy is predicted if biopsy specimens show predominant lymphocytes uncontaminated with other types of leucocytes (10). Seventy- five percent of pseudotumor cases have demonstrated positive response to radiotherapy (10). In cases where steroid or radiotherapy is not the suitable options, second- line immune- modulating agents may be useful. Histological evidence should be obtained in cases which do not respond to initial steroid treatment to confirm the diagnosis and direct towards better treatment.

Conclusion

Orbital inflammatory pseudotumor is a relatively common inflammatory disease of the orbit, yet it proves to be a diagnostic challenge. Optic nerve sheath meningioma may mimic the orbital pseudotumour in radiological imaging, if other orbital structures involvement is not apparent. Radiotherapy offers an alternative treatment options, yet recurrences is a possibility. Treatment with steroid is effective in majority of cases. Biopsy is without doubt a necessity to confirm the diagnosis, yet it may not always be amenable in all cases.

References

- Jacobs D, Galetta S. Diagnosis and management of orbital pseudotumor. *Curr Opin Ophthalmol* 2002; 13(6):347-51.
- Uehara F, Ohba N. Diagnostic imaging in patients with orbital cellulitis and inflammatory pseudotumor. *Int Ophthalmol Clin* 2002;42(1):133-42.
- Karesh JW, Baer JC, Hemady RK. Noninfectious orbital inflammatory disease. In: Tasman W, Jaeger EA, eds. *Duane's Clinical Ophthalmology*. Philadelphia: Lippincott Williams & Wilkins, 2005, pp-1- 45.
- Badr AM, Elkhamary SM, Al Sabangh M, Al Turjoman AA. Bilateral optic nerve sheath meningioma with intracanalicular and intracranial component in a 25 year-old Saudi patient. *Middle East Afr J Ophthalmol* 2008;15(3):138-41.
- Chaudhry IA, Shamsi FA, Phil M, Arat YO, Riley F. Orbital pseudotumor: distinct diagnostic features and management. *Middle East Afr J Ophthalmol* 2008;15(1):17-27.
- Kapur R, Sepahdari AR, Mafee MF, et al. MR imaging of orbital inflammatory syndrome, orbital cellulitis and orbital lymphoid lesions: the role of diffusion- weighted imaging. *AJNR Am J Neuroradiol* 2009;30(1):64-70.
- Gordon LK. Orbital inflammatory disease: a diagnostic and therapeutic challenge. *Eye* 2006; 20(10): 1196- 1206.
- Rubin PA, Foster CS. Etiology and management of idiopathic orbital inflammation. *Am J Ophthalmol* 2004;138(6):1041-3.
- Notter M, Kern T, Forrer A, Meister F, Schwegler N. Radiotherapy of pseudotumor orbitae. *Front Radiat Ther Oncol* 1997; 30: 180–191.
- Orcutt JC, Garner A, Henk JM, Wright JE. Treatment of idiopathic inflammatory orbital pseudotumors by radiotherapy. *Br J Ophthalmol* 1983;67(9):570-4.