Orbital pseudotumor : A conservative management
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Abstract
A Sixty five years old female reported to eye OPD of Centre for Woman and Child Health (CWCH), Baron, Ashulia, Dhaka. On 27/5/2008, with the complaint of headache, unilateral proptosis in her left eye, swelling of the left eyelid for 3 years, congestion of the conjunctiva and chemosis of the left lid. Nasal pterygium of left eye. Cornea was clear, pupil was reacting to light. No abnormalities were detected on anterior chamber. No history of vomiting, no loss of vision and her vision was 6/6V6/6, she was suffering from pansinusitis. Ophthalmoscopic examination on both eyes revealed no abnormality. On slit lamp examination right eye was normal in size and shape. On left eye there was a swelling in the medial canthus which shifted the eye ball towards left side on looking forward; eye ball deviated towards left about less then 45 degree from the coronal plain. Her x-ray chest showed no abnormality. All haematological parameters were found within normal range. CT scan of the orbits, PNS & brain done. On orbits isodense intraconal soft tissue density oblong fashion mass was seen on left orbit, which shows moderate heterogeneous enhancement after contrast introduction. No evidence of erosion or sclerosis of bone adjacent to mass. The mass compressed & closely adheres to sclera but may not invaded to intrabulbar space. Mild proptosis of left eyeball. Right orbit appears to be unremarkable. In Brain- no evidence of intra or extra axial hemotoma, contusion, mass (especially in the visual pathway), infarct, abnormality in gray-white matter interface or in the appearance of sulci & gyri. Ventricles and basal cisterns are unremarkable. Impression says a soft tissue intraconal mass in left orbit, possibilities were inflammatory pseudotumor, hemangiona, schwannoma and dermoid. Although the incidence of orbital pseudotumor is low in our country. The aim of the article is to raise the awareness among the eye practitioners about the disease and its management1.

Keyword
Orbital pseudotumor, proptosis.

Introduction
The orbit is a bony, pyramid-shaped cavity in the skull. It contains and protects the eyeball and related structures. The eyeball lies in the anterior part of the orbit, enclosed in a facial sheath which separates it from the orbital muscles and fat2. Orbital pseudotumor is a benign idiopathic inflammatory process involving the structures in the orbit which may present in several different patterns. It may be acute, subacute, chronic or is rare cases recurrent. It may diffusely involve the entire orbit or present in a localized fashion involving either the anterior orbit, posterior orbit, lacrimal gland or extraocular muscles. Most cases are unilateral, but bilateral cases are seen and are more common in children.

Orbital pseudotumor is a swelling of the orbital tissues behind the eye, but unlike cancerous tumors, it cannot invade other tissues or spread elsewhere. The cause is unknown. It most commonly affects young women, although it can still occur at any age. Pain on eye movement, decreased vision, eyelid swelling, red eye (rare). Most cases are mild and do well. Severe cases may be resistant to treatment and visual loss may occur. Orbital pseudotumor usually involves only one eye. Severe cases of orbital pseudotumor may push the eye forward to the extent that the lids can no longer protect the cornea, leading to drying of the affected eye.

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This can lead to damage to the clarity of the cornea or to corneal ulcer. The eye muscles may not be able to properly aim the eye and double vision may result. The changes of pseudotumor can be seen when the eye is examined. Tests to differentiate a pseudotumor from a tumor include the following: ultrasound, skull x-ray, biopsy. There is no racial or gender predilection with orbital inflammatory pseudotumor (OIP) and patients may range from 4 to 80 years. While chronic forms do exist, the typical presentation includes acute onset of orbital pain, swelling, chemosis and proptosis. Orbital pseudotumor may be associated vision loss with concurrent disc edema or optic atrophy secondary to nerve compression & often ophthalmoplegia and diplopia. The disease is typically unilateral and may be recurrent. However, this condition may be commonly bilateral in children. The term is used to describe any idiopathic inflammatory lesion that simulates a neoplasm within the orbit. It presents with compressive effects to orbital structures with evidence of inflammation and infiltration. It is believed to be a self-limiting disease and while benign in nature, can cause serious ocular damage and possibly vision loss from optic nerve compression.

Orbital inflammatory pseudotumor can be subdivided into different types: granulomatous, sclerosing, vasculitic and eosinophilic, depending upon the histological characteristics. Granulomatous orbital pseudotumor is characterized by histiocytes and multinucleate giant cells. Sclerosing orbital pseudotumor presents with minimal inflammatory infiltrate with a greater degree of interstitial connective tissue. Vasculitic inflammatory pseudotumor involves primary vasculitis of small vessels with lymphocytes and granulocytes destroying the muscularis and elastic lamina of the vessels. Eosinophilic pseudotumor involves tissue eosinophilia without vasculitis. Mechanisms ranging from autoimmunity to infectious to poor wound healing have all been proposed to account for the development of orbital inflammatory pseudotumor. The end effect is an inflammatory infiltrate occupying space and compressing tissues, vessels and nerves with a mass-lesion effect similar to a true tumour.

There are strong similarities between orbital inflammatory pseudotumor (OIP) and the Tolosa-Hunt syndrome of painful ophthalmoplegia, which is rare condition caused by non-specific granulomatous inflammation of the cavernous sinus, superior orbital fissure and orbital apex, signs are proptosis, ocular motor nerve palsies, often with involvement of the pupil sensory loss along the distribution of the first and 2nd division of trigeminal nerve. Diagnostic neuro-radiological testing of these two diseases shows identical signal intensity, though it is in different locations. Orbital inflammatory pseudotumor is typically orbital and Tolosa-Hunt syndrome is predominately retro-orbital, specifically localizing to the anterior cavernous sinus. This, in combination with nearly identical clinical presentations and histopathologic findings make the two diseases nearly indistinguishable. Some have theorized that they are on the continuum of the same condition. High-resolution CT scan will demonstrate soft tissue swelling, but this is not the diagnostic modality of choice. Oral corticosteroids are the recommended treatment for OIP. This condition is extremely steroid-responsive and was, at one time, considered a diagnostic finding. Oral prednisone 60mg to 80mg qd with rapid tapering upon clinical improvement is acceptable therapy.

Discussion
The orbital pseudotumor is defined as non-specific, non-neoplastic inflammatory process of the orbit without identifiable local/systemic cause. The disorder was first described by Birch-Hirschfield in 1905. This is a diagnosis of exclusion based on history, the clinical course of the disease and the response to steroid therapy, laboratory tests and biopsy in a limited number of cases. There is a group of disease entities that can mimic pseudotumors such as lymphoid tumors, thyroid orbitopathy, sarcoïdosis, and other granulomatous diseases.
Case Report

Each of these abnormalities, or at least components of each, has been included under the umbrella term pseudotumor at sometime during the last few decades. Currently the term pseudotumor should be reserved for idiopathic orbital inflammatory syndrome.

Idiopathic orbital inflammatory syndrome accounts for 4.7% to 6.3% of orbital disorders and the disease is more prevalent in adults than pediatric population. The pathogenesis of the disease remains elusive but several lines of evidence point to immune mediated processes as the likely underlying mechanism. Orbital pseudotumor may have protean clinical manifestations. The most common being unilateral, sudden onset ocular pain, proptosis and impaired/loss of vision. Some presentation of idiopathic orbital inflammatory syndrome may mimic conditions such as orbital cellulites. Idiopathic orbital inflammatory syndrome is usually confined to orbit, rarely may extend intracranially. However, when confined to orbit, may be multifocal, involving more than one structure. Most authors recognize it as, (a) myositis, (b) dacrycystadenitis, (c) periscleritis, (d) perineuritis, or (e) diffuse group. The diffuse group refers to the patients in whom involvement of orbital fat predominates. It is the diffuse variety which simulates lymphomatous infiltration and biopsy is helpful in establishing the diagnosis in such cases. In the subgroup of perineuritis, there is inflammation of the sheath surrounding the optic nerve. In addition to idiopathic orbital inflammatory syndrome, optic neuropathy may be seen in number of conditions such as multiple sclerosis, optic nerve sheath meningioma, autoimmune disease (systemic lupus erythematosus), post viral infections (herpes, chicken pox, rubella, etc.) and infective conditions as syphilis, toxoplasmosis, Lyme disease. Radiation optic neuropathy is another rare cause. It is important to diagnose the underlying cause of the optic neuritis since prognosis and treatment varies for each condition, acute pain is an important feature of pseudotumors. Multiple sclerosis usually presents with gradual painless loss of vision. Immune markers with other systemic presentation would negate the diagnosis of pseudotumors. Viral neuritis is usually seen after 10-14 days after the primary illness. The index case was found negative for antinuclear antibodies and there was no history of preceding illness. Since patient presented with acute pain with impaired vision without any systemic involvement, hence a diagnosis of optic neuritis or pseudotumor was considered. CT may show some enlargement of optic nerve, usually with some degree of enhancement. MR imaging will depict the thickening of the optic nerve. There may be few streaky densities in contiguous orbital fat. Post contrast fat-suppressed, T1-weighted MR images may be the best technique to demonstrate optic neuritis as seen in index case. Contrast enhancement is often subtle or present in short segment of nerve, particularly in intracanalicular portion of the nerve. Steroids show a dramatic effect in the treatment of acute cases of pseudotumors and usually reverse the changes completely.

Case report

A sixty five year's old female reported to eye OPD with the complaint of headache, unilateral proptosis in her left eye, swelling of the left eyelid for 3 years, congestion of the conjunctiva, chemosis of the left lid. Nasal pterygium of left eye. Cornea clear, pupil is reacting to light. No abnormality were detected on anterior chamber. No history of vomiting, no loss of vision, her vision was 6/6V6/6, she was suffering from sinusitis. Ophthalmoscopic examination on both eyes revealed no abnormality. On slit lamp examination right eye was normal in size and shape. On left eye there was a swelling in the medial canthus which shifted the eyeball towards left side on looking forward; eyeball deviated towards left about more then 45 degree from the coronal plain. "Joffroy's sign-absence of wrinkling of the forehead on
Case Report

Looking upwards with the face inclined downwards was positive. Her x-ray chest showed no abnormality. All haematological parameters were found within normal range. CT scan of the orbits, PNS & brain: On orbits isodense intraconal soft tissue density oblong fashion mass was seen at left orbit, which showed moderate heterogeneous enhancement after contrast introduction. No evidence of erosion or sclerosis of bone adjacent to mass was found. The mass compressed & closely adheres to sclera but did not invade to intrabulbar space. Mild proptosis of left eyeball was seen. Right orbit appears to be unremarkable. PNS- mucosal thickening is noticed at bilateral maxillary, ethmoidal, sphenoidal & frontal sinuses and hypertrophied left nasal turbinates with narrowing of ipsilateral nasal passage. Brain- no evidence of intra or extra axial hematoma, contusion, mass (especially in the visual pathway), infarct, abnormality in gray-white matter interface or in the appearance of sulci & gyri ventricles and basal cisterns are unremarkable. No evidence of contra lateral shifting of mid-line structures was seen. Pituitary region- no intra, para or suprasellar mass was seen, posterior fossa- no lesion at brain stem, cerebellum or at CP angle was seen. No mass/ lesion is noticed at initial course of the cranial nerves. Impression- soft tissue intraconal mass at left orbit, possibilities are inflammatory pseudotumor, hemangioma, schwannoma, dermoid. Clinico-pathological correlation is suggested for further evaluation. Normal CT finding of the brain. But about paranasal sinus impression shwos pansinusities. Her x-ray chest P/A view revealed no abnormality. Blood report- haemoglobin (Hb) 0.62%, total WBC- 9000 per c.mm. Different WBC count- neutrophil-60%, lymphocyte-34%, monocyte- 02%, eosinophil-04%, basophil-00%. E.S.R-20mm in 1st hour. Blood sugar random-4.95 mmol/l, hormone test- T3-0.98ng/ml and T4-9.66ug/dl (Impression- serum level of T3 & T4 are within normal range). FSH - 72.82 mlU/ml. ECG within normal limit.

Treatment

Patient was treated with oral prednisolone 900 mg/day (Tablet 60mg) in divided doses and it was tapered down with 60 mg per day within 30 days, with under cover of omeprazole 20 mg bid doses. With addition NSAID was given for 3 days bid.

After two week the swelling become just half. Full dose steroid therapy was completed. After one month, swelling reduced to almost normal size. But eye ball was mildly deviated laterally from its normal position. But her vision was intact.

Conclusion

Whenever any patient present to an ophthalmologist with complains of headache, vomiting, swelling and defective vision. He or she should be thoroughly examined & investigated like visual acuity, slit lamp examination, intraocular pressure (IOP), fundoscopy direct and indirect both. If there is any intraorbital swelling, ophthalmologist should ask for CT or MRI. Most of the time orbital pseudotumor present with disturbed vision but in this case it was not. So ophthalmologist should refer the case to radiologist with full clinical notes for CT or MRI. So the radiologist can help the ophthalmologist in diagnosis the diseases type, extension, localization & extension of ocular lesion.

References

Case Report