CASE REPORT

Bilateral sclerosing orbital pseudotumour in an adult

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SUMMARY

Sclerosing pseudotumour of the orbit is a rare idiopathic chronic inflammatory process, which greatly mimics neoplasms both clinically and on imaging studies. It is therefore important to differentiate this entity from true neoplasms and to rule out any systemic associations. We present a rare case of sclerosing orbital pseudotumour in an adult man with no systemic illnesses who presented with painless progressive proptosis of both eyes and showed a gradual initial response to steroid therapy but relapsed after a 2-year interval.

BACKGROUND

Bilateral orbital pseudotumour in the absence of a systemic association is rare and is a diagnosis of exclusion. Systemic associations such as lymphoproliferative disorders in adults and rhabdomyosarcoma, leukaemia, neuroblastoma, etc in children need to be ruled out before a diagnosis of orbital pseudotumour can be performed. Identifying the aggressive nature of this entity and differentiating it from neoplasms becomes crucial so that aggressive medical management can be initiated and unnecessary surgical intervention avoided.

CASE PRESENTATION

A 58-year-old man presented to the radiology department with a history of gradually progressive, painless, bilateral proptosis since 6 months. The proptosis was greater on the left side with associated chemosis. The proptosis had increased in severity in the past 2 months and was associated with watering of eyes, burning sensation and diplopia. There was also restricted movement of the extraocular muscles on both sides, more so on the left. Visual acuity was decreased with 6/36 vision in both eyes; however, there was no palpable mass on either side. Fundus examination revealed bilateral optic disc oedema and congested veins. No history of flashes or floaters or ocular trauma was present. There was no history of thyroid disease, diabetes, hypertension or other comorbidities.

INVESTIGATIONS

Blood tests were performed as part of the workup and revealed normal haemoglobin level, red blood cell (RBC) and white cell counts (WCCs) and thyroid function. ESR was elevated at 58 mm/h. Peripheral smear showed normocytic normochromic RBCs and adequate number of WCCs and platelets.

Contrast-enhanced CT of the orbits showed minimally enhancing retrobulbar masses filling and expanding the intraconal space on both sides (figure 1A,B). Extraocular muscles were normal.

The optic nerves, however, were not separately visualised. There was no underlying bone destruction or intracranial extension. Based on the above CT findings, a differential diagnosis of lymphomatous infiltration, orbital pseudotumour and granulomatous disease was considered.

MRI of the orbit showed an intraconal mass which was hypointense on T2-weighted (T2W), isohypointense on T1-weighted (T1W: figure 2A–C) and enhanced heterogeneously on postcontrast T1W sequence (figure 2D). Optic nerves were encased by the mass; however, extraocular muscles were normal in signal intensity. There was no intracranial extension.

Biopsy of the lesion was performed and revealed foamy histiocytes, lymphocytic aggregates, plasma cells, few neutrophils and occasional giant cells intermixed with proliferating spindle cells of fibroblastic/myofibroblastic type in a sclerotic stroma with rich vasculature. A histological diagnosis of sclerosing orbital pseudotumour was performed.

TREATMENT

The patient was started on oral (tablet wysolone 30 mg/day) and topical steroids (methycellulose drops) and antibiotics. There was no significant reduction in the proptosis for nearly 3 weeks post-treatment. Steroid treatment, however, was continued and there was gradual improvement in the patients' symptoms with a decrease in proptosis and improvement of visual acuity. He was discharged and advised a follow-up examination after 6 months.

OUTCOME AND FOLLOW-UP

At the end of 6 months, he reported of further reduction in visual acuity and visually evoked potential test showed prolongation of the p100 latency suggesting bilateral optic nerve involvement. Steroid treatment was continued. The patient then came back only after a period of 2 years with recurrence of proptosis which was more in the left eye. A repeat imaging was advised to look for the present status of disease and the patient was offered a more aggressive line of therapy. However, he refused any further intervention.

DISCUSSION

Orbital pseudotumour is an idiopathic, non-neoplastic inflammatory condition of the orbit, which was first described in the year 1905 by Birsch-Hirschfeld.^{1–3} It is the third most common disease of the orbit after Grave's ophthalmopathy and lymphoproliferative disorders constituting approximately 8–11% of cases.⁴ It is seen commonly in children and young adults; however, it

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Figure 1 (A and B) Axial and coronal postcontrast CT sections (Philips Brilliance 64 slice CT scanner using 60 mL of intravenous contrast, omnipaque—300 mgI/mL) showing minimally enhancing retrobulbar mass in the intraconal compartment bilaterally. There is no extraocular muscle involvement. No bone destruction seen.

does not seem to have any gender predilection. Bilateral pseudotumour in adults in the absence of systemic disease is uncommon. It is mainly a diagnosis of exclusion and association with a systemic disorder must be ruled out with the help of history, clinical examination and imaging. Orbital pseudotumour presents with a broad spectrum of clinical manifestations and may take an aggressive course which many times is refractory to standard therapy and hence requires more aggressive treatment. However, identifying the true nature of this condition and

recognising it from other malignant pathologies is important to avoid unnecessary surgical intervention. Histological confirmation with biopsy may be required when there is a poor response to steroid therapy. Recurrences are also known to occur in this condition.

Clinical findings

Clinically it may present with periorbital oedema, erythema, proptosis, ptosis, diplopia or painful eye movements.⁵

Figure 2 (A–C) Axial T1-weighted (T1W), T2-weighted (T2W) and coronal STIR images (GE 1.5 T scanner, T1—TE-9 TR-360, T2—TE-103 TR-4400, STIR—TE-83 TR-6280) showing bilateral retrobulbar lesion (arrows) which are hypointense on T1W and hypointense on T2W and STIR sequence. The optic nerves (arrowheads) are encased by the mass. No intracranial extension was seen. (D) T1W postcontrast sequence showing significant contrast enhancement. STIR, short tau inversion recovery.

Involvement is usually unilateral. Children usually present with bilateral disease and no associated systemic cause.

Imaging findings

Imaging modalities used for diagnosis are CT and MRI; however, serological tests may be required to exclude a systemic cause. On CT and MRI there may be a diffuse mass involving the retrobulbar tissues, infiltration of the intraconal fat, orbital inflammation, uveoscleral thickening, contrast enhancement of tenon's space and thickening of extraocular muscles and optic nerve.^{6,7} Inflammation of extraocular muscle or myositis can be acute, subacute or recurrent with involvement of the medial rectus being common. Here there is an involvement of the entire muscle belly including its tendinous attachment. This can be differentiated from Grave's disease where there is fusiform enlargement of muscle with sparing of the tendon insertion.⁸ All these changes including infiltration of adjacent fat, periocular involvement or discrete mass with thickened muscles help in making a diagnosis of pseudotumour.

MRI plays an important role in the diagnosis of sclerosing orbital pseudotumour, especially the T2W sequence. The most typical imaging feature of fibrosing/sclerosing inflammatory pseudotumour on MRI is a soft-tissue mass which is hypointense relative to the brain on T1W and T2W images showing homogenous postcontrast enhancement. In acute cases there is T2W hyperintensity due to acute inflammation and oedema. However, as the disease becomes chronic there is increase in the amount of fibrosis and these lesions become hypointense on T2W.⁹ Hypointensity of these lesions on T2W images clinches the diagnosis and is probably due to the decreased free water content, associated lack of mobile protons related to a high degree of fibrosis and also due to an elevated cellularity or high nucleus-to-cytoplasm ratio.¹⁰

This entity is closely related to benign retroperitoneal fibrosis, sclerosing mediastinitis, fibrotic thyroiditis, etc and shows poor response to steroids and radiotherapy and warrants aggressive immunotherapy.^{11,12}

Whenever there are bilateral orbital pseudotumours involving the retrobulbar region and the apex, it is suggested that systemic causes be ruled out and a biopsy be performed. This is to rule out sclerosing orbital pseudotumour which is a difficult disease to diagnose on imaging and manage as is seen in our case. Sclerosing pseudotumour is a chronic slow-growing process involving the orbital structures making it difficult to differentiate from the neoplasm.¹³ This form of orbital pseudotumour is a distinct disease entity which requires aggressive intervention as it responds poorly to corticosteroids. A second line of treatment using chemotherapy or radiotherapy may have to be used. Recurrences are especially common in bilateral orbital pseudotumours.

Bilateral orbital pseudotumour is more common in children as compared with adults and is seen in 45% of cases. Some of the common differential diagnoses in children include orbital cellulitis, rhabdomyosarcoma, leukaemia, neuroblastoma, etc.¹⁴ There is a known association of orbital pseudotumour with ocular and systemic disorders which include scleritis, rheumatoid arthritis, Crohn's disease and systemic lupus erythematosus.^{2,3} It may also be associated with Wegener's granulomatosis, fibrosing mediastinitis, autoimmune thyroiditis and sclerosing cholangitis.

Extraorbital extension of orbital pseudotumour may occur and may contiguously involve adjacent structures such as the cavernous sinus, conal structures, maxillary sinus and the anterior or middle cranial fossa.¹⁵ Intracranial extension of orbital

pseudotumour is rare and occurs through superior orbital fissure into the middle cranial fossa and cavernous sinus. It mimics a neoplastic or infective process and is treated with high-dose corticosteroids with the use of radiotherapy in refractory cases.¹⁶ Isolated involvement of the optic nerve by pseudotumour in patients presenting with gradual progressive loss of vision is also rare and responds dramatically to corticosteroids.¹⁷

In approximately one-third of cases, it is difficult to distinguish between pseudotumour and lymphoma on histopathological examination and immunophenotypic studies and genetic analyses are required to make a diagnosis.¹⁸

Treatment

Corticosteroids are the treatment of choice with 75% of patients improving dramatically in 24–48 h.^{2,3} In patients who are resistant to steroids, radiotherapy may be helpful. A pseudotumour in its acute form is more responsive to high doses of systemically administered prednisone than a chronic lesion as was seen in our case.¹⁹ A recent case report on intraorbital inflammation highlights the dramatic response to steroids.²⁰ In patients who respond neither to corticosteroids nor radiation, chemotherapeutic agents such as cyclophosphamide, methotrexate and cyclosporine have been found to be helpful.²¹

Learning points

- ▶ Bilateral orbital masses in adults should include a differential diagnosis of orbital pseudotumour, lymphoma and Grave's disease.
- ▶ Sclerosing orbital pseudotumour is a rare entity which requires aggressive intervention and responds poorly to steroids. Hence its correct diagnosis is of utmost importance.
- ▶ MRI shows a hypointense soft tissue mass on T1-weighted and T2-weighted (T2W) sequences which shows homogenous postcontrast enhancement.
- ▶ Hypointensity of these lesions on T2W sequence clinches the diagnosis which is due to decreased free water content and associated lack of mobile protons related to a high degree of fibrosis.

Competing interests None.

Patient consent Obtained.

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