Computed Tomography (CT) of Orbital Pseudotumour

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Summary
Orbital pseudotumour is a non-specific inflammatory disease. Its highly variable clinical and radiographic presentation makes specific diagnosis difficult. This study on 31 proven cases of pseudotumour shows that unilateral involvement, retro-orbital fat infiltration and enlargement of multiple extraocular muscles with involvement of the tendinous insertions are key computed tomography CT features which help to establish the diagnosis of orbital pseudotumour.

Key words: Orbit, pseudotumour, computed tomography (CT).

Introduction
Orbital pseudotumour is a non-specific orbital inflammatory process of unknown etiology that may affect part or all of the tissue within the orbit. It has a broad range of clinical presentation and many of the symptoms are non-specific. Its varied presentation is due to the different sites of involvement of the pseudotumour within the orbit and, as such, even its radiographic appearance is varied. This makes CT diagnosis difficult as well. CT appearance can mimic thyroid ophthalmopathy or any infiltrative disease such as orbital lymphoma. This study on 31 proven cases of orbital pseudotumour is to evaluate the spectrum of CT characteristics and to determine key CT findings which help to establish the diagnosis of pseudotumour.

Materials and Methods
The CT findings of documented orbital pseudotumour in 31 patients presenting with proptosis between January 1987 to December 1990 were reviewed. There were 17 males and 14 females between the ages of 4 to 75 years. The diagnosis of orbital pseudotumour was based on the clinical history, clinical course, response to steroid and muscle biopsy which was performed on 8 patients.

All patients had axial CT with either 4.5 mm sectional thickness, contiguous slices using Phillips Tomoscan 350 or 5 mm sectional thickness, contiguous slices using Toshiba TCT-900s. Additional coronal scans with either 4.5 mm or 6 mm contiguous slices using Phillips Tomoscan 350 or 5 mm using Toshiba TCT-900s were
done on 12 patients. The coronal sections were helpful in identifying the optic nerve, the superior rectus and inferior rectus muscles. Intravenous contrast was given to 18 patients where there was suspicion of a vascular lesion or suspicion of an orbital mass with possible intracranial extension.

Results

The CT features analysed were lateralisation of involvement, retroorbital fat infiltration, extraocular muscle enlargement, muscle tendon involvement, lacrimal gland infiltration, orbital bone remodelling and sinusitis (Table I). Four patients (13%) had bilateral involvement, making a total of 35 pseudotumours available for analysis. The most common intraorbital abnormality was retroorbital fat infiltration, followed by enlargement of extraocular muscles and thickening of muscle tendons.

Retroorbital fat infiltration was seen as areas of increased attenuation within the retroorbital fat, causing the adjacent margins of recti muscles and optic nerve to be indistinct. Of the 29 orbits which had retroorbital fat infiltration, the distribution of the infiltration was patchy in 14 orbits (48%). Nine orbits (31%) showed infiltration confined to the posterior aspect of the globe and 3 orbits (10%) showed infiltration confined to the apex. Diffuse retroorbital fat infiltration was seen in 3 orbits (10%). Of the 23 orbits with extraocular muscle enlargement, there was multiple muscle involvement in 15 (65%). Sixteen orbits (70%) showed thickening of the muscle tendon (the region of muscle insertion onto the globe). A total of 59 extraocular muscles were affected: 18 (31%) lateral rectus, 15 (25%) medial rectus, 13 (22%) superior rectus-levator complex and 13 (22%) inferior rectus.

Discussion

The manifestation of pseudotumour on CT is variable. Its appearance has been classified according to its anatomic involvement into 5 patterns, namely: myositic, anterior, posterior, diffuse and lacrimal. In this study, there were 19 (54%) myositic, 7 (20%) anterior, 3 (8.5%) posterior, 3 (8.5%) diffuse and 3 (8.5%) lacrimal types.
Fig 1: Myositis pseudotumour involving the left orbit. There is enlargement of medial and lateral recti muscles extending anteriorly to involve the tendinous insertions to the globe (arrowheads).

Fig 2: Anterior pseudotumour of the right orbit. The posterior wall of the globe is thickened (arrowheads). Infiltration extends posteriorly along the optic nerve (arrow).
In earlier reports, the CT appearances of orbital myositis have been described as a typically unilateral process involving a single extraocular muscle. A subsequent study, however, reported multiple muscle involvement, which is also a common occurrence in our study. The distribution of involvement of myositis in our series revealed the lateral rectus to be the most commonly involved muscle, followed by the medial, superior rectus-levator complex and the inferior rectus. In orbital myositis, extraocular muscle enlargement extends anteriorly to involve the tendinous insertion onto the globe. This ‘tendon sign’ is regarded as a reliable diagnostic indicator of orbital myositis and was seen in 70% of orbits in our study (Fig 1).

Extraocular muscle enlargement is by no means a feature confined to orbital pseudotumour as it is seen in other conditions which include thyroid ophthalmopathy, carotico-cavernous fistula, cavernous sinus-dural arteriovenous malformation, metastases and infiltrative disease. The typical appearance of thyroid ophthalmopathy is different from orbital myositis. It is usually bilateral, although it can be asymmetrical. In this condition, the muscle enlargement is typically fusiform due to enlargement of the muscle belly with sparing of the tendinous insertions to the globe. The muscles commonly involved are the inferior rectus followed by the medial, superior and lateral recti muscles. There is no infiltration of the retroorbital fat. Hence, the margins of the extraocular muscles and the optic nerve are sharply defined. Carotico-cavernous fistula and cavernous-dural arteriovenous malformation are associated with enlargement of the superior ophthalmic vein and fullness of the ipsilateral cavernous sinus. Extraocular muscle enlargement from direct tumour invasion or metastasis may be accompanied by adjacent infiltrative orbital mass, nodularity or irregular enhancement of the muscle.

In the anterior type of pseudotumour, there is infiltration of retroocular fat at the posterior aspect of the globe (Fig 2). The infiltration extends posteriorly along the optic nerve and there is thickening of the posterior wall of the globe.

In the posterior type of pseudotumour there is infiltration of fat at the apex of the orbit obscuring the margins of the muscles and optic nerve (Fig 3). The infiltration extends anteriorly along the recti muscles and optic nerve.

**Fig 3:** Posterior pseudotumour of the right orbit. Apical infiltration extends anteriorly along the medial rectus (arrowhead) and the optic nerve (arrow). There is patchy infiltration of the retroorbital fat of the left orbit.
Fig 4: Diffuse pseudotumour of the right orbit. The entire retrobulbar space is filled by a hyperdense soft tissue mass moulding itself around the posterior aspect of the globe (arrowheads). The lateral rectus muscle and tendon are enlarged (arrow). The optic nerve is seen as a central lucent band within the mass.

Fig 5: Lacrimal pseudotumour of the left orbit. The lacrimal gland is enlarged (arrowheads). Infiltration extends along the posterior wall of the globe which is thickened (arrow).
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In diffuse pseudotumour the entire retrobulbar space is filled with a dense soft tissue mass extending from the posterior margin of the globe to the orbital apex (Fig 4). The mass moulds itself around the globe without distorting the shape of the globe. This type of pseudotumour is difficult to differentiate from lymphoma. Lymphoma is less likely to cause muscle enlargement with tendinous involvement. However, biopsy would be necessary for definitive diagnosis. True orbital tumours such as optic nerve glioma or meningioma will indent and distort the posterior margin of the globe.

The lacrimal type of pseudotumour is confined to the superolateral aspect of the orbit with enlargement of the lacrimal gland and infiltration into the lateral aspect of the globe and lateral rectus (Fig 5).

Conclusion

In orbital pseudotumour, CT is valuable in defining the area of orbital involvement, the extent of infiltration and response to steroid. However, because of its varied appearances, it is difficult to make a CT diagnosis of orbital pseudotumour with absolute certainty. The majority of cases of orbital pseudotumour in our series are the myositic type. Unilateral involvement, enlargement of the tendinous insertions and retroorbital fat infiltration are the key CT indicators of orbital pseudotumour.

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References