Radiology Quiz

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Clinical History

Middle age patient (male) presenting a rapid development of a left unilateral axile painful exophthalmos, associated with swollen lids and vessel engorgement on orbital clinical inspection and examination. Blurred vision from the left eye is reported. A slight limitation of vertical eye movements is observed. The orbital auscultation is normal. Moderate venous engorgement on the ocular fundus is retained. A history of remitting course with spontaneous regression of the proptosis or after steroid treatment is elicited.

Figure 1a - Proton density weighted axial image.

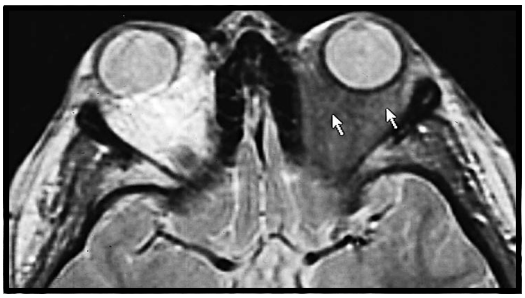

Figure 1b - Proton density weighted axial image.

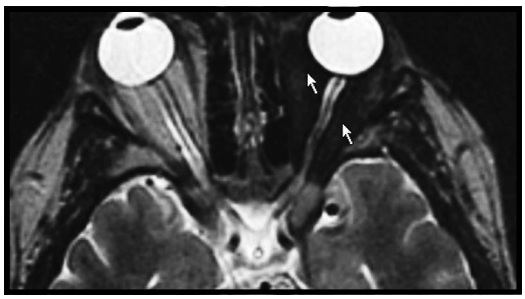

Figure 2a - T2 weighted axial image.

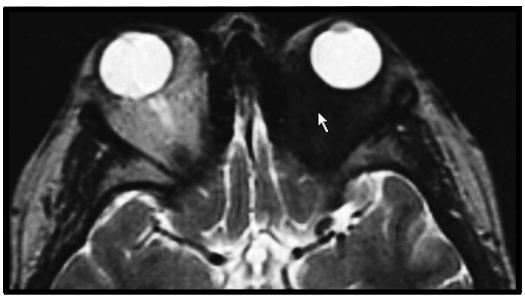

Figure 2b - T2 weighted axial image.

Questions

1. What are the radiographic abnormalities?
2. What is the diagnosis?

Answer Page

Figure 1a - Proton density weighted axial image.

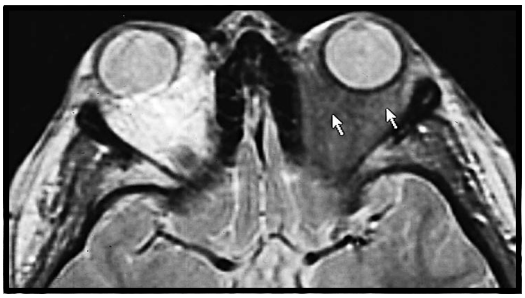

Figure 1b - Proton density weighted axial image.

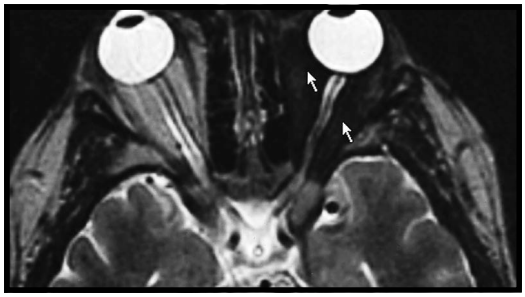

Figure 2a - T2 weighted axial image.

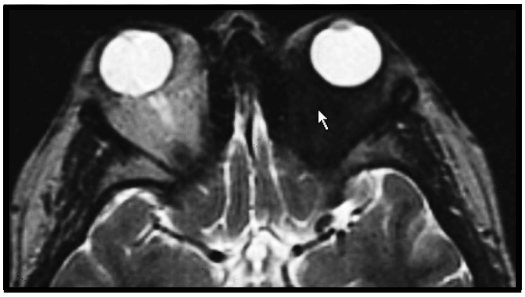

Figure 2b - T2 weighted axial image.

Findings and Discussion

Question 1 - Mass infiltrating the retrobulbar and extraconal spaces, presenting a relative hyposignal on T1W and a hypointense signal on heavily T2W, surrounding the intraorbital optic nerve and sheaths. Note the axile exophthalmos. The lesion showed enhancement after Gadolinium infusion (not shown). The hypointense on T2W probably relates to the presence of desmoplastic fibrosis. Note that some malignant orbital lesions such as lymphoma and myeloma may show similar signal patterns to pseudotumor.

Question 2 - **Inflammatory orbital pseudotumor**

Clinical and pathological features: It is a non specific idiopathic process, most commonly unilateral, in adults. Most commonly acute: pain, proptosis, ocular motor disturbance with diplopia. May be chronic: orbital mass syndrome. One of the most frequent causes of unilateral exophthalmos (apart from Basedow's). Many clinical forms: scleritis, myositis, dacryoadenitis, orbital fat infiltrate. Special entity: Tolosa-Hunt syndrome. If bilateral: rule out systemic diseases such as, collagen-vascular, Wegener, trichinella, or lymphoid. Dramatic response in acute forms to high dose steroid therapy.

Magnetic resonance characteristic findings: *1. Morphology*: Various forms: restricted or diffuse infiltrate. Irregular uveal-scleral thickening. Orbital muscle enlargement involving typically the tendinous insertion. Isolated lacrimal gland enlargement (frequent). Ill-defined infiltration of the orbital fat "dirty orbit". Perioptic involvement similar to lymphoproliferative diseases. Sclerosing pseudotumor subtype: in chonic form or as an end stage of the acute form, showing a diffuse mass "freezing the intra-orbital structures". Tolosa-Hunt subtype: involvement of the orbital apex and the cavernous sinus. *2. Signal intensity*: Shows some specificity, due to fibro-collagenous composition. SE-T1w: isointense to orbital muscles. SE-T2w: isointense or hypointense to muscles and orbital fat. Lymphoma, myeloma, sarcoidosis or Grave's diseases may mimic pseudotumor. Usually different SI behavior from metastatic carcinoma (hyperintense on T2). *3. Contrast enhancement:* Diffuse orbital or focal (muscle, sclera, lacrymal gland, optic nerve sheaths, or cavernous sinus) enhancement.

References

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2. Jones IS, Jakobiec F. Diseases of the Orbit. Paris, Berlin, Heidelberg, New York: Harper and Row Publishers; 1979. p. 620.