Case 1051

Eurorad

Tolosa-Hunt syndrome

Published on 06.05.2001

DOI: 10.1594/EURORAD/CASE.1051
ISSN: 1563-4086
Section: Neuroradiology
Imaging Technique: MR
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Case Type: Clinical Cases
Authors: S. Cakirer(1), D. Cakirer(2), G.M. Galip(1)
Patient: 34 years, male

Clinical History:

A 34 year-old male patient with headache, nausea, vomiting, left sided orbital-periorbital pain, followed by left sided oculomotor and abducens nerve palsies, ptosis, and miosis. These findings regressed with corticosteroid treatment.
Imaging Findings:

A 34 year-old male patient had an episode of headache, nausea, vomiting, left sided orbital-periorbital pain for a period of 4 days, followed by left sided oculomotor and abducens nerve palsies, ptosis, and miosis within 3 days of the beginning of the symptoms. The serological tests supported an inflammation, 100 mg/day methyl prednisolone treatment was started at the 4th day. Clinical and serological findings regressed, but not totally resolved within two weeks. An MRI examinaton of the cranium was performed with SE T1 and FSE T2 weighted, and post-gadolinium SE T1 weighted sequences on sagittal, axial, coronal planes at the beginning of the treatment.
Discussion:

Tolosa-Hunt syndrome (THS) is characterized by painful ophthalmoplegia due to idiopathic granulomatous inflammation of the cavernous sinus. Steroid therapy dramatically reverses the clinical signs and symptoms of THS. The International Headache Society defined the diagnostic criteria of THS in 1988 to include at least one episode of unilateral or bilateral orbital pain for an average of 8 weeks if untreated, with associated paresis of one or more of the third, fourth, and sixth cranial nerves, and exclusion of other causative lesions by neuroimaging and (not compulsory) carotid angiogram. Cranial nerve paresis may coincide with the onset of pain or follow it within a period of up to 2 weeks, and the pain must be relieved within 72 h after the initiation of corticosteroid therapy. The patients with THS may have some nonspecific systemic symptoms, such as back pain, chronic fatigue, arthralgia, gut problems. Seventy % of the patients show positive serological findings for inflammation during an attack of THS. MRI shows a unilateral enlargement of the symptomatic cavernous sinus by an abnormal tissue isointense with gray matter on T1weighted images and iso-hypointense on T2 weighted images. This abnormal tissue markedly enhances after intravenous contrast injection. It may extend into the contiguous regions, mainly the orbital apex and subtemporal fossa ipsilaterally. In the presence of painful ophthalmoplegia, the finding by MRI of cavernous sinus enlargement, with the herein described signal and extension characteristics and slow resolution with corticosteroid treatment, is highly suggestive of the Tolosa-Hunt syndrome. However other causative lesions must be excluded by neuroimaging, especially of the region of the cavernous sinus and the orbita, and by blood and CSF examinations. Since imaging techniques have dramatically improved, it is now possible to visualize the inflammatory tissue in THS. MRI findings help in the differential diagnosis of the Tolosa-Hunt syndrome from conditions such as meningioma, lymphoma, sarcoidosis, and tuberculosis, as well as confirming the similarities of the Tolosa-Hunt syndrome and orbital pseudotumor. Some authers propose that THS is probably related to orbital pseudotumor due to a possible communication of orbital and cavernous sinus inflammation through the superior orbital fissure. Orbital pseudotumor

responds to corticosteroid treatment like THS.
Differential Diagnosis List: Tolosa-Hunt syndrome

Final Diagnosis: Tolosa-Hunt syndrome

References:

Hannerz J. Recurrent Tolosa-Hunt syndrome: a report of ten new cases. Cephalalgia 1999 Dec;19 Suppl 25:33-5. (PMID: 10668117)
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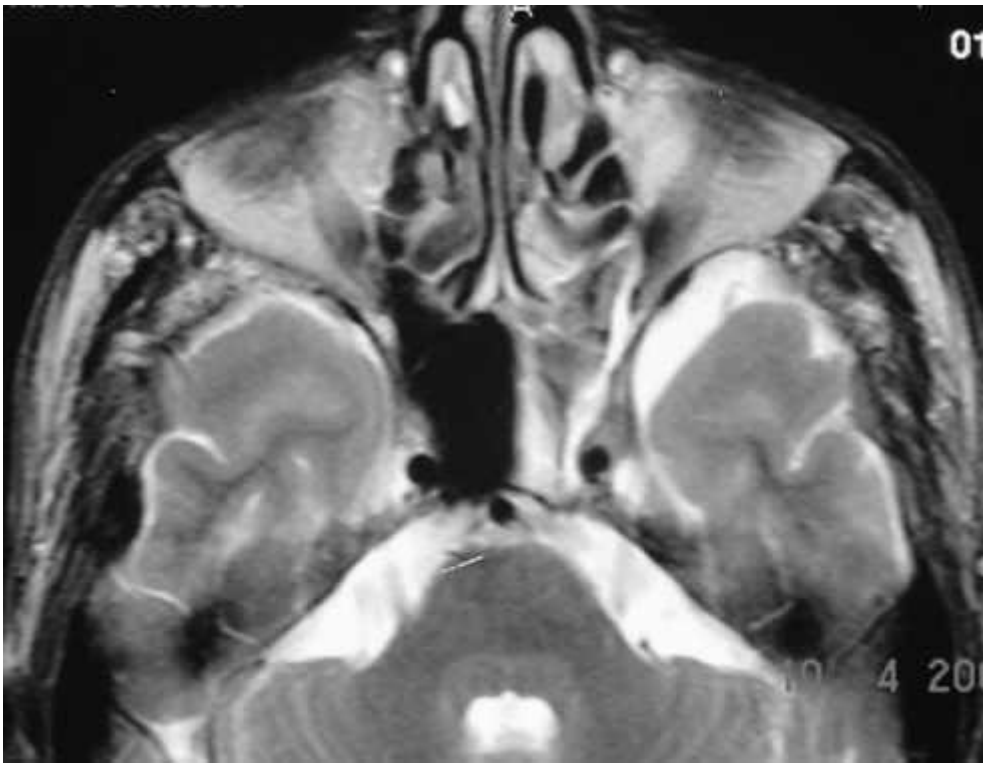
Alioglu Z, Akbas A, Sari A, Erdol H, Ozmenoglu M. Tolosa Hunt syndrome: a case report. Clinical and magnetic resonance imaging findings. J Neuroradiol 1999 Mar;26(1):68-72. (PMID: 10363446)

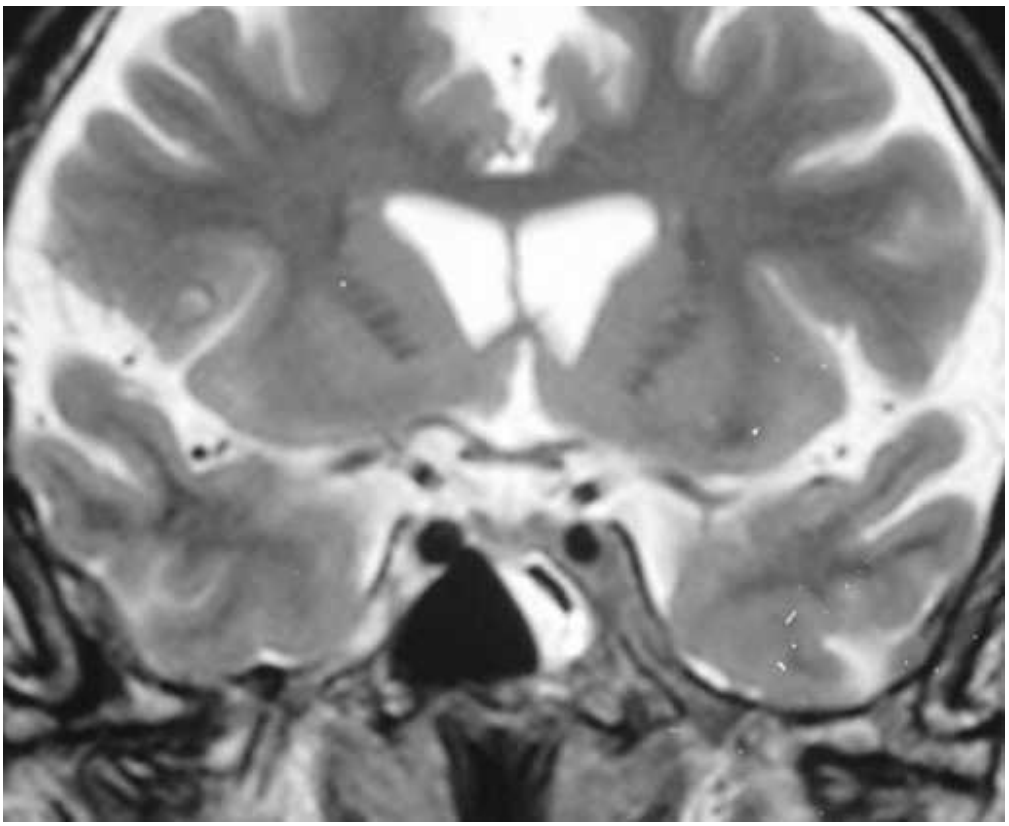
Figure 1

a



Description: Axial FSE T2-weighted MR image demonstrates widening of the left cavernous sinus with a homogeneous tissue mildly hypointense to gray matter. Note that there is a focal collection on the left side of the sphenoidal sinus. **Origin:**

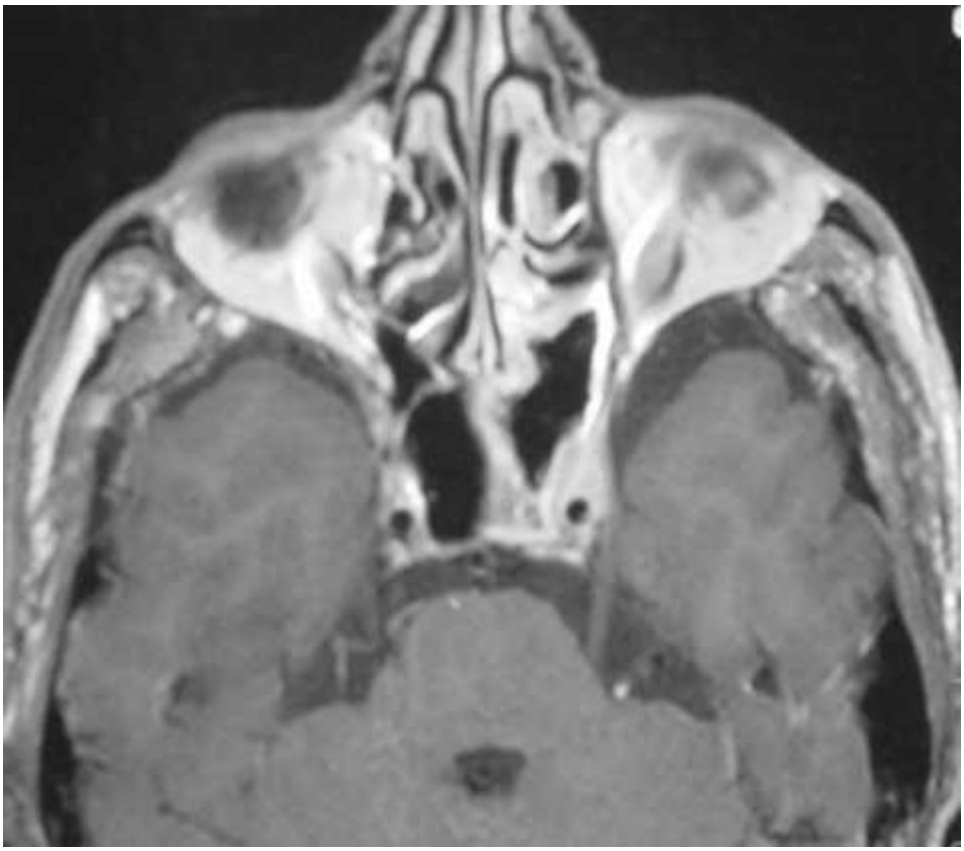
b



Description: Coronal FSE T2-weighted MR image clearly delineates widened left cavernous sinus with a homogeneous tissue mildly hypointense to gray matter. **Origin:**

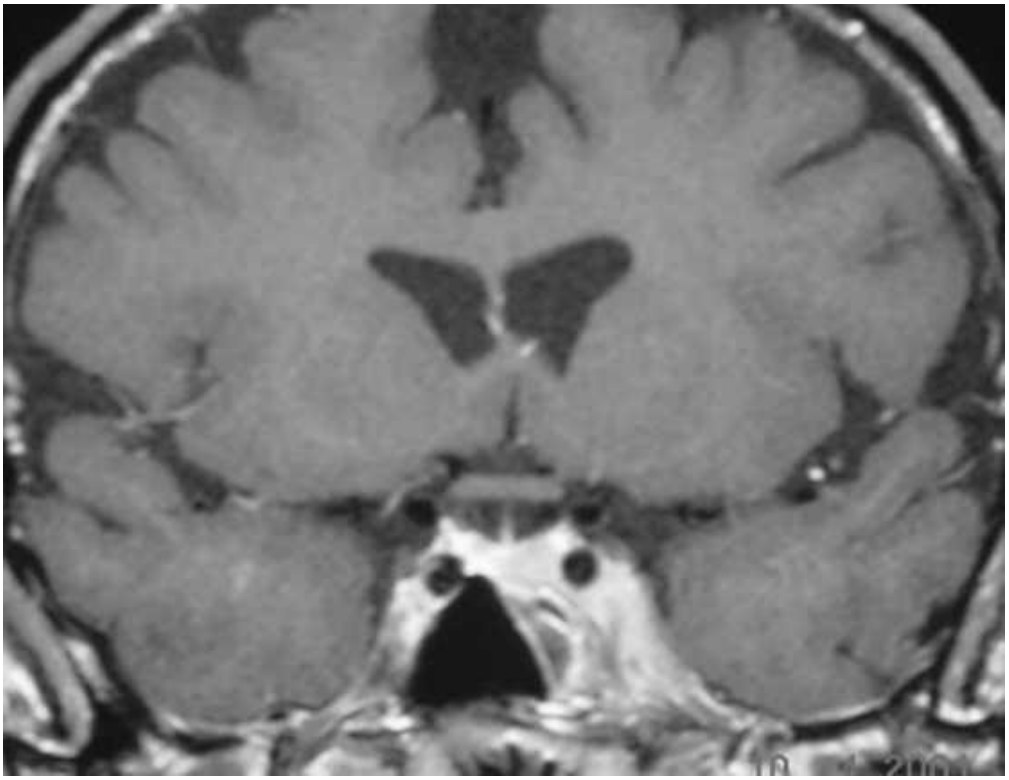
Figure 2

a



Description: Axial SE T1-weighted MR image following intravenous contrast injection reveals an intense contrast enhancement of the thickened parasellar tissue. **Origin:**

b



Description: Coronal SE T1-weighted MR image following intravenous contrast injection reveals an intense contrast enhancement of the thickened parasellar tissue. **Origin:**