Tolosa Hunt Syndrome: A Case Report

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Abstract

Tolosa-Hunt syndrome (THS) is a painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure. We present a case of THS disease evaluated on a 1.5 Tesla MR scanner.

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A fifty-year-old man presented with painful diminution of vision and diplopia in left eye developing over a period of 7 days. There was tingling sensation over left half of upper face along with the above-mentioned complaints.

Based on the imaging findings a diagnosis of THS was made and the patient was started on corticosteroid therapy: injection methylprednisolone (1gm.i.v for 3 days). There was dramatic improvement in diplopia and pain within 48 hours of institution of therapy. After 3 days oral prednisolone 40mg. OD was started and continued for 6 weeks. At the end of 6 weeks the patient’s ophthalmoplegia also recovered.

Discussion

The clinical differential diagnosis of steroid responsive painful ophthalmoplegia includes metastases, carotid-
cavernous fistulae, pituitary adenomas, vasculopathic cranial neuropathy, aspergillus invasion, Wegener's granulomatosis, sarcoidosis, lymphoma and ophthalmoplegic migraines are not associated with masses in the cavernous sinus or orbital apex as in THS.

Fig. 3: Coronal T1wi: An Isointense Soft Tissue In Left Cavernous Sinus

Fig. 4: Post Gado T1wi: Enhancing Soft Tissue In The Left Cavernous Sinus And Orbital Apex

Meningiomas and aneurysms may rarely cause pain when of sufficient size. While metastases, pituitary adenomas, aspergillus infection, some meningiomas and some cases of lymphoma are often hyperintense relative to fat on long TR images, sarcoidosis, lymphoma and meningiomas may display hypointensity or isointensity on short TR/TE and long TR/TE sequences as in THS. However sarcoidosis and lymphoma will often have systemic symptoms and meningiomas will not resolve with steroid therapy. Vascular abnormalities such as arteritides, cavernous-carotid fistulae, ophthalmoplegic migraines and aneurysms are not associated with masses in the cavernous sinus or orbital apex as in THS.

Fig 5: Coronal Post Gado Image

Fig 6: Palsy of left internal oblique

Fig 7: Palsy of left inf. rectus
Tolosa Hunt Syndrome

THS is caused by an inflammatory process first described by Tolosa[2] as granulomatous periarteritis of cavernous carotid. Hunt et al[3] described proliferation of fibroblasts and infiltration of septa and wall of cavernous sinus with lymphocytes and plasma cells. Its cause is unknown. Pathologically this syndrome falls within the range of idiopathic orbital pseudotumor.[4]

Administration of systemic steroids for 48 hours in a patient with THS produces a dramatic response in painful ophthalmoplegia that allows differentiation of this cause from other conditions of painful ophthalmoplegia.[5]

The abnormal area in the cavernous sinus in THS is of intermediate intensity on T1W1. This is consistent with the pathological process of THS that is granulomatous inflammation[6]. In the appropriate clinical setting of painful ophthalmoplegia, MR findings of a cavernous sinus abnormality that is isointense with muscle on short TR/TE images and hypointense to isointense with fat on long TR/TE images suggests the diagnosis of THS.

REFERENCES

Amersham