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.

On clinical examination, there was left III, IV and VI CN palsy, chemosis in left eye and decreased sensation over the area supplied by V1 CN. The patient was sent for an MR examination.

MR imaging was performed on a 1.5 Tesla MR scanner (Sonata, Siemens). 10-ml. Gadolinium was injected in the left antecubital vein to obtain post contrast T1WI. The study revealed moderately enhancing soft tissue in the region of left cavernous sinus and orbital apex. It was isointense to muscle on T1WI and hypointense to fat on T2WI (TSE).

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 migraine.

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.[1] 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.
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 pseuotumor.[4]

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