

Tolosa Hunt Syndrome- A Case Report

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Summary:

Tolosa Hunt syndrome is a rare disorder of uncertain aetiology characterized by severe periorbital headaches and recurrent painful unilateral ophthalmoplegia. Resolution of findings on follow-up imaging and response to steroids is characteristic.

This case is to emphasize the role of MRI in the diagnosis of this condition.

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Introduction:

Tolosa Hunt syndrome is a rare disorder characterized by severe periorbital headaches and recurrent painful unilateral ophthalmoplegia. The major symptoms include chronic periorbital headache, double vision, certain cranial nerve palsies and chronic fatigue. Affected individuals may also exhibit protrusion of eyeball, drooping of upper eyelids and diminished vision.

It is caused by non specific inflammation of cavernous sinus or superior orbital fissure, which is responsive to steroid therapy [1]. The cause of the constant pain, which characterizes the onset of the disorder, is due to infiltration of lymphocytes and plasma cells along with thickening of dura matter within the cavernous sinus.

The exact cause of Tolosa Hunt syndrome is unknown. One theory is an abnormal autoimmune response linked with an inflammation in a specific area behind the eye (cavernous sinus and superior orbital fissure). Other possible causes may include generalized inflammation and constricted or inflamed cranial blood vessels.

Condition may be sight threatening if untreated inflammation extends beyond cavernous sinus to affect optic nerve.

Our case report is regarding a 26 years old male presenting with left orbital pain and restriction of motility

of left eye in all gazes. The clinical presentation can be a pointer to several conditions of the cavernous sinus and a correct diagnosis is a must to institute appropriate early management.

This case is to emphasize the role of MRI in the diagnosis of this condition.

Case Report:

Twenty six years old male presented with complaints of severe, left sided unilateral headache and blurring of vision 30 days and drooping of left eyelid for 10 days. Headache was limited to left fronto-temporal region.

Ocular motility of left eye was restricted in all gazes.

Ocular movements on the right side was normal.

Conjunctiva was congested.

No sensory loss over face was detected. Motor system was normal.

There was no associated fever, vertigo or arthralgia.

Patient was diagnosed as Total External Ophthalmoplegia Left Eye with involvement of 3rd, 4th and Upper 2 divisions of 5th cranial nerve.

Patient underwent extensive laboratory workup, which was nonspecific.

Patient was referred for a CT brain which showed, enhancing lesion measuring approx. 2.7 cm in AP dimension along left cavernous sinus, extending upto left optic foramen.

Subsequent contrast enhanced MRI examination of brain and orbit was performed on the same day in a SIEMENS 3T. 0.1mmol/kg Gadolinium was administered.

Pre and post contrast Coronal and Axial sequences of the cavernous sinus were done. Additionally FS CE T1W

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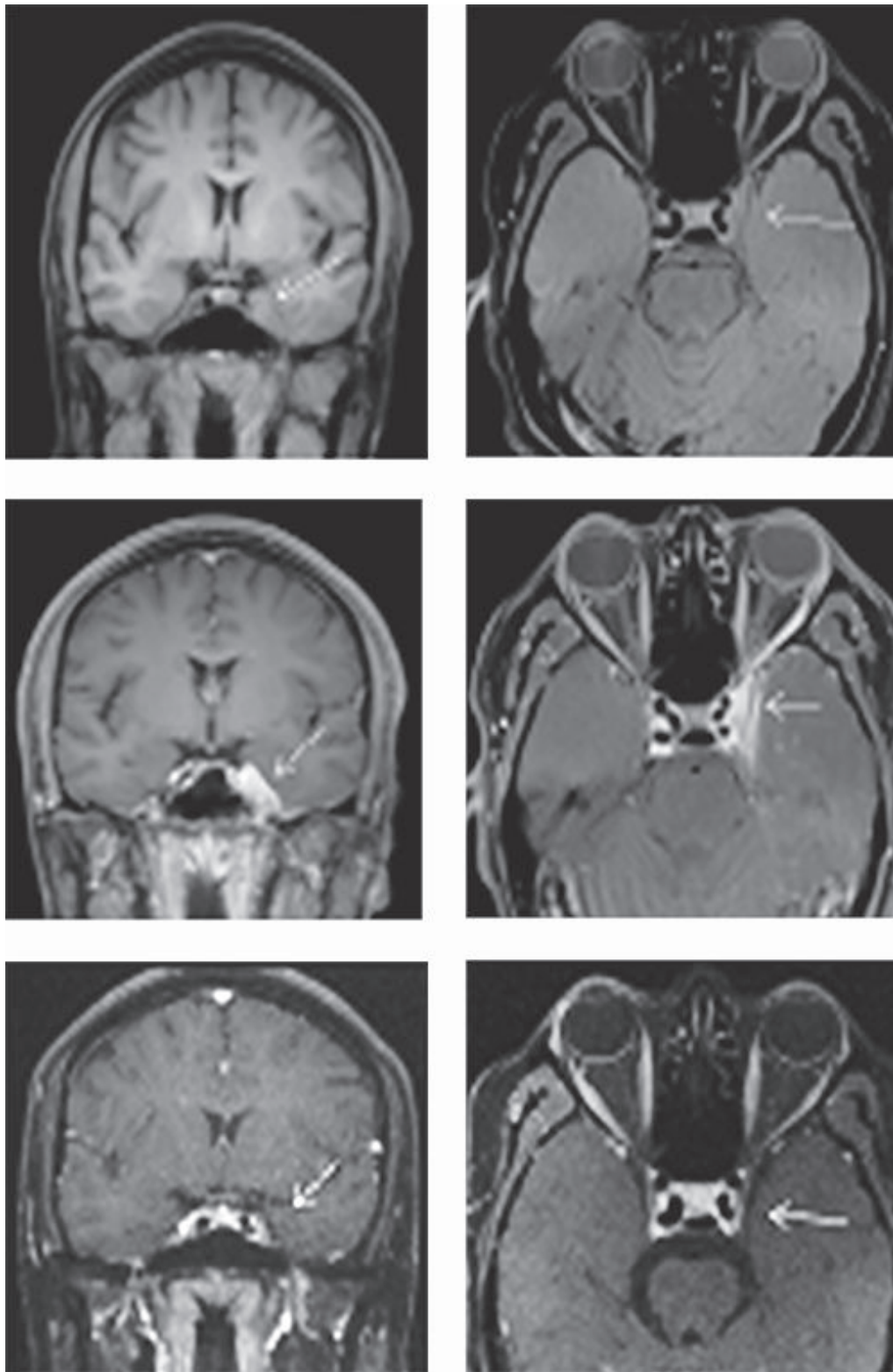


Fig.-1: Precontrast T1W Coronal and axial MRI images, CE MRI images and Follow up MRI images.

sequences were also done.

CE MRI showed expansion of cavernous sinus by an enhancing soft tissue mass, at left parasellar region, measuring approx. 27 mm (AP) x 9 mm (TR) in size, extending anteriorly upto left optic foramen and posteriorly traced upto left trigeminal nerve.

No luminal narrowing of the internal carotid artery was demonstrated.

3D TOF MRA and 2D TOF MRV revealed no additional abnormalities. The rest of the study was unremarkable.

The above imaging findings were consistent with Tolosa hunt syndrome within the context of clinical history. Differentials were Sarcoidosis, Lymphoma.

Patient was treated with steroids (Tab Cortan 40 mg bd). There was dramatic relief of symptoms subsequently. Follow-up demonstrated improvement in diplopia and ocular movement came back to normal.

Further periodic follow up was done and patient is doing well.

Follow up MRI usually shows complete resolution of abnormality.

Discussion:

First described by Tolosa in 1954 in a patient with unilateral recurrent painful ophthalmoplegia with involvement of cranial nerves III, IV, and VI. Carotid angiogram showed narrowing of carotid siphon. HUNT described similar features in 6 patients in 1961. The entity was termed Tolosa Hunt syndrome by

Smith and Taxdol (1966).

THS is essentially a clinical diagnosis of exclusion. Exclusion of other conditions by neuroimaging is important. Pain that is relieved within 48 hrs of steroid therapy is characteristic. Before CT the radiographic

evaluation consisted of angiography and plain films to exclude aneurysm, meningioma, metastases and pituitary masses. Angiography include narrowing of carotid siphons, occlusion of superior ophthalmic vein, non-visualization of cavernous sinus. However, normal orbital venogram/arteriogram does not exclude THS 2.

Disease manifests as recurrent attacks of steady, dull retro orbital pain, palsies of third, fourth or 6th cranial nerves and first or second divisions of V cranial nerve and venous engorgement. Pathologically there is infiltration of lymphocytes and plasma cells along with thickening of duramater.

Conclusion:

Tolosa Hunt syndrome is essentially a diagnosis of exclusion. The role of radiologist is to exclude other conditions causing similar clinical features. Resolution of findings on follow-up imaging and response to steroids is characteristic.

MRI seems to be the ideal technique to follow progressive resolution of the abnormal tissue after steroids 3.

CE MRI plays a vital role in the early diagnosis of this sight threatening disorder 4.

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