

Case Report

Tolosa-Hunt syndrome: a rare case report with uncommon imaging findings and discussion

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ABSTRACT

A 45 year old diabetic but non hypertensive female presented with unilateral ptosis and complete external ophthalmoplegia on the left side. All the routine investigations were inconclusive. A signal void change was found in MRI study including angiogram of brain. On performing VEP (visually evoked potential), there was mild left optic pathway dysfunction (axonal and demyelinating). A probable diagnosis of Tolosa-Hunt Syndrome (THS) was made and the patient was started on steroids. She responded dramatically to the therapy and was discharged on steroids. Thus the diagnosis of THS was confirmed. On follow up after one month, both her ptosis and ophthalmoplegia had resolved completely. On subsequent follow up visits, she was free of any ophthalmological symptoms and signs.

Keywords: Tolosa-Hunt syndrome, Ophthalmoplegia, Steroids

INTRODUCTION

Tolosa-Hunt Syndrome (THS) is a rare disorder indicated by recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or Superior Orbital Fissure (SOF). The disease shares histopathological features with idiopathic orbital pseudotumour; however, owing to its anatomical location, it produces characteristic clinical manifestations.¹

Recurrent retro-orbital pain, with palsies of the third, fourth or sixth cranial nerves as well as the first and second divisions of the trigeminal nerve, are typical. Clinically, immediate response to steroid therapy is a hallmark of the condition. The clinical presentation of THS has a wide differential diagnosis, and timely and appropriate imaging - as an adjunct to pertinent laboratory investigations - can greatly assist clinicians with early accurate diagnosis and management.

CASE REPORT

Our patient, a 45 years old known diabetic, but non hypertensive lady presented with a 4 week history of left hemicranial headache and drooping of the left eyelid. She also complained of pain, redness, swelling, protrusion, watering from left eye for the same duration. Diplopia, maximum on the left side and blurring of vision in the same eye was also present.

On clinical examination her vitals were normal, she had left sided ptosis, left sided sluggish pupillary reflex and restriction of movement of left eye in all direction. All extraocular muscles were involved, (left 3rd, 4th, 6th cranial nerves) (Figure 1 & 2). Visual acuity was intact. There was no sensory or motor loss in the distribution of trigeminal nerve (5th). Fundoscopy was normal. No other cranial nerve involvement could be elicited. Sensory and motor system examination was normal. No long tract signs were present. Bladder and bowel habits were within normal limits. No other abnormalities were found.



Figure 1: Presenting features before steroid therapy showing left sided ptosis.

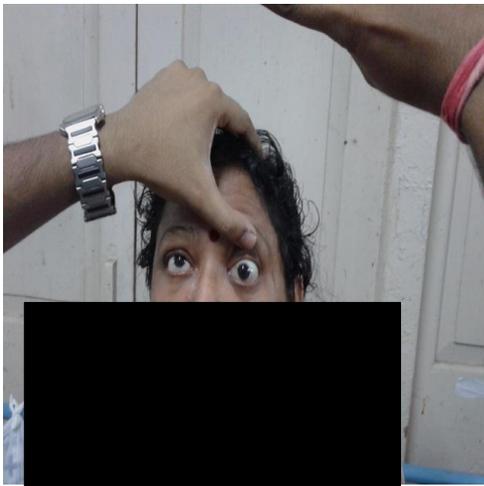


Figure 2: Presenting features before steroid therapy showing left sided ophthalmoplegia.

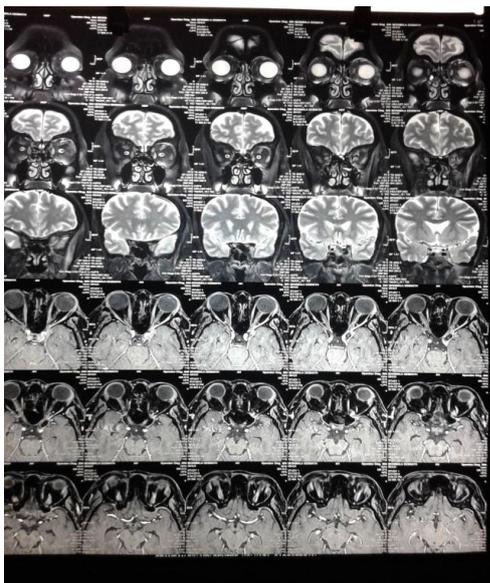


Figure 3: MRI brain.

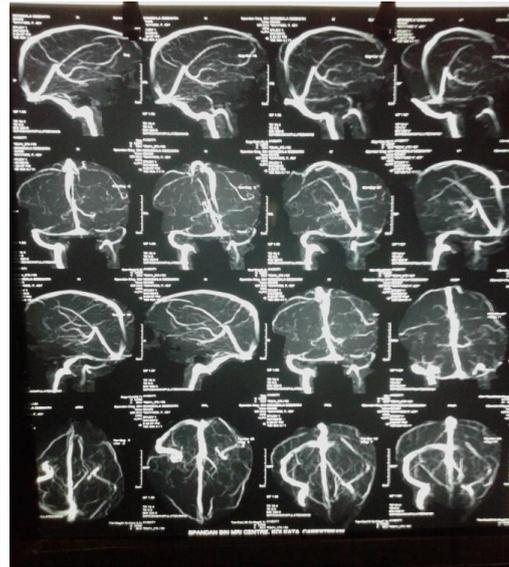


Figure 4: MR venogram brain.

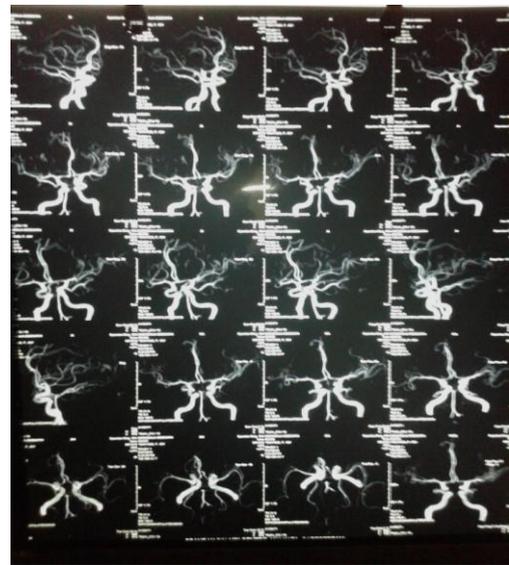


Figure 5: MR arteriogram brain.



Figure 6: Same patient after 1 month steroid therapy showing resolution of ptosis.



Figure 7: Same patient after 1 month steroid therapy showing resolution of ophthalmoplegia.

Complete hemogram and Lumbar Puncture (LP) were noncontributory. Hb% was slightly decreased and ESR slightly raised. HbA_{1C} was 7.4%. Viral markers for retrovirus (HIV I/II) and hepatitis virus were negative. Tuberculosis and syphilis were also ruled out.

CT scan brain was normal. A contrast MRI of brain and orbits was then performed - again no abnormality was detected. Orbit and ocular muscles did not show any abnormal signal enhancement (Figure 3). MR Angiogram of the brain was also normal (Figure 4 & 5). A small signal void area was seen in left transverse sinus, most likely due to a flow void signal because of turbulent effect. On performing VEP (visually evoked potential), there was mild left optic pathway dysfunction (axonal and demyelinating). AEP/BAER (auditory evoked potential brain AEP) were normal in both ears.

Based on these features and tests, a probable diagnosis of Tolosa-Hunt syndrome was made. The patient was treated with oral steroids and showed significant relief of symptoms dramatically over the following 48 hours, thus confirming the diagnosis made earlier. She was discharged on oral steroids which were gradually tapered off. She had complete resolution of the ptosis as well as ocular nerve palsies within one month. Further follow up showed no recurrence of symptoms and signs.

DISCUSSION

Tolosa first described the condition in 1954, in a patient with unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, VI and V1. The patient was imaged using carotid angiography, and segmental narrowing of the carotid siphon was observed.¹ Hunt et al. described 6 patients with similar clinical findings in 1961, and proposed a low-grade non-specific inflammation of the cavernous sinus and its walls as the plausible cause of the syndrome. Pathophysiologically, infiltration of lymphocytes and plasma cells as well as thickening of the dura mater was noted.¹ the condition

was named Tolosa-Hunt syndrome by Smith and Taxdal in 1966.² The latter authors also emphasized the importance of the dramatic and rapid response to steroid therapy. In 1988, THS criteria were provided by the International Headache Society (IHS), and further amended in 2004.

Diagnostic criteria^{3,4}

- A. One or more episodes of unilateral orbital pain persisting for weeks if untreated.
- B. Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or demonstration of granuloma by MRI or biopsy.
- C. Paresis coincides with the onset of pain or follows it within 2 weeks.
- D. Pain and paresis resolve within 72 hours when treated adequately with corticosteroids.
- E. Other causes have been excluded by appropriate investigations.

Neuro-imaging - in particular MRI - is an essential part of the workup of any patient presenting with features of THS, as these features are non-specific and have a wide range of differential diagnosis, including meningioma, ophthalmoplegic "migraine", sarcoidosis, pituitary tumours, tuberculous meningitis (TBM) and lymphoma. MRI findings classically show a soft-tissue mass lesion involving the superior orbital fissure or cavernous sinus. Signal characteristics are typically hypointense to fat and isointense to muscle on short TR/TE sequences and isointense to fat on long TR/TE sequences.⁵ Significant enhancement of the mass lesion is demonstrated on contrast enhanced sequences. Of particular value are the post-contrast fat-saturated thin-slice coronal images through the orbital apex and cavernous sinus.⁶ THS essentially remains a diagnosis of exclusion. The patient described above did not show the classical MRI findings and was diagnosed on the basis of exclusion and response to steroid therapy. The role of the radiologist is to exclude other conditions causing similar clinical features. Distinctive MRI findings and rapid resolution of clinical symptoms with steroid therapy are characteristic.

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Ethical approval: Not required

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