

A Rare Case of Tolosa Hunt

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INTRODUCTION

Tolosa-Hunt Syndrome is a rare clinical entity, the exact pathology of which is still unknown. THS is caused by a non-specific inflammatory process in the cavernous sinus or superior orbital fissure (SOF). Although painful ophthalmoplegia is not rare, THS as a cause of painful ophthalmoplegia has been considered as a rare clinical entity. The etiopathogenesis of THS largely remains unknown. No exact information is available on what actually triggers the acute inflammatory process within the cavernous sinus/superior oblique fissure. Thus, the syndrome can be taken as a manifestation of idiopathic orbital inflammation (Pseudotumor).^[7] The syndrome of painful ophthalmoplegia consists of periorbital or hemicranial pain combined with ipsilateral ocular motor nerve palsies, oculosympathetic paralysis, and sensory loss in the distribution of the ophthalmic and occasionally the maxillary division of the trigeminal nerve. Various combinations of these cranial nerve palsies may occur, localizing the pathological process to the region of the cavernous sinus/superior orbital fissure. We are presenting a case report on THS. The patient presented with complete ptosis, unilateral headache and diminished vision. On examination, there was complete ophthalmoplegia and diminished sensation over the area supplied by 1st and 2nd division of trigeminal nerve. CEMRI brain revealed enhancing soft tissue thickening along right cavernous sinus extending to right orbital apex without any e/o filling defect in cavernous sinus or dilated SOV suggestive of THS. Other differential diagnoses were excluded on the basis of careful history, examination and investigations.

In 1966, Smith and Taxdal applied the eponym "Tolosa-Hunt Syndrome".^[1] Tolosa-Hunt Syndrome is a rare clinical entity with annual incidence 1 per million worldwide^[2] which is characterized by unilateral headache or retro orbital pain with palsy of multiple cranial nerves mainly 2nd, 3rd, 4th, first division of 5th and occasionally second division of 5th cranial nerve with significant improvement with steroids. In most cases, aetiology is unknown. In 1954, Tolosa^[3] reported the first patient with this syndrome, who presented with left orbital pain, ipsilateral progressive visual loss, total left ophthalmoplegia, and reduced sensation over the first division of the trigeminal nerve. After 7 years of this, in 1961, Hunt et al.^[4] described this clinical entity, on the basis of six patients. It is an idiopathic condition which develops due to non-specific granulomatous inflammatory process in the region of the cavernous sinus/superior orbital fissure.

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- 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 granulomas 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.

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PRESENTATION OF CASE

A 60-years-old woman presented to the medicine OPD with sudden onset of right sided headache since 7 days followed by drooping of the right upper eyelid since 5 days. The headache was sudden in onset, throbbing in nature involving the right half of head and around the right eye, extending to frontal and temporal region and not associated with nausea and vomiting. After 2 days, patient developed ptosis of right eye. Patient was non-diabetic and non-hypertensive. There was no history of head trauma. On examination, the patient was afebrile, with a pulse rate of 82 beats/min and blood pressure of 130/80 mmHg. There were no signs of pallor, icterus, cyanosis, clubbing, raised jugular venous pressure, or pedal oedema. She was conscious and oriented and other higher mental functions were normal. On ocular examination, there was complete ptosis on the right side at the time of presentation. On eye examination visual acuity of both eye is 6/9. Pupil was non-reactive to light. Medial rectus palsy of right eye is present on examination. Patient is unable to lift his right sided eye lid.

Examination of other cranial nerves was normal. Other system examination was essentially normal. Both direct and indirect pupillary reflexes were absent on right side initially but with start of steroid therapy, pupil became mid dilated and started sluggishly reacting to light.

platelet count 1.6 lac. Blood sugar was 96 mg/dL with normal liver and renal function test. Her ESR was 06 mm. Human immunodeficiency virus testing by enzyme-linked immunosorbent assay was negative. ANA testing was found to be negative. Cerebrospinal fluid (CSF) routine and microscopic finding is within normal limit. ECG, X-ray chest, and ultrasound abdomen revealed no abnormal findings.

Moderate enhancing mild soft tissue thickening seen involving anterior part of bilateral cavernous sinuses and bilateral superior orbital fissure right more than left likely acute inflammatory aetiology? Acute stage of Tolosa Hunt syndrome.

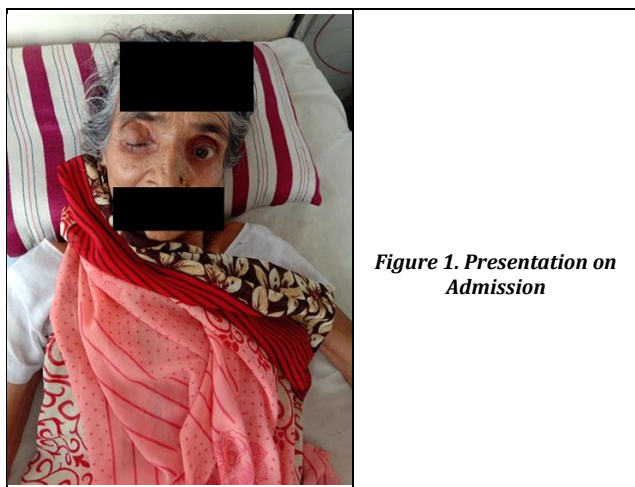


Figure 1. Presentation on Admission

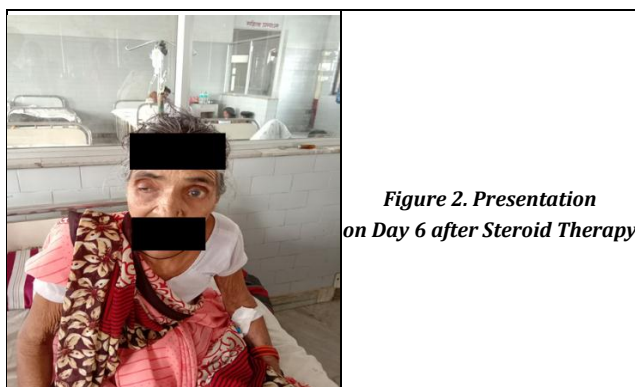
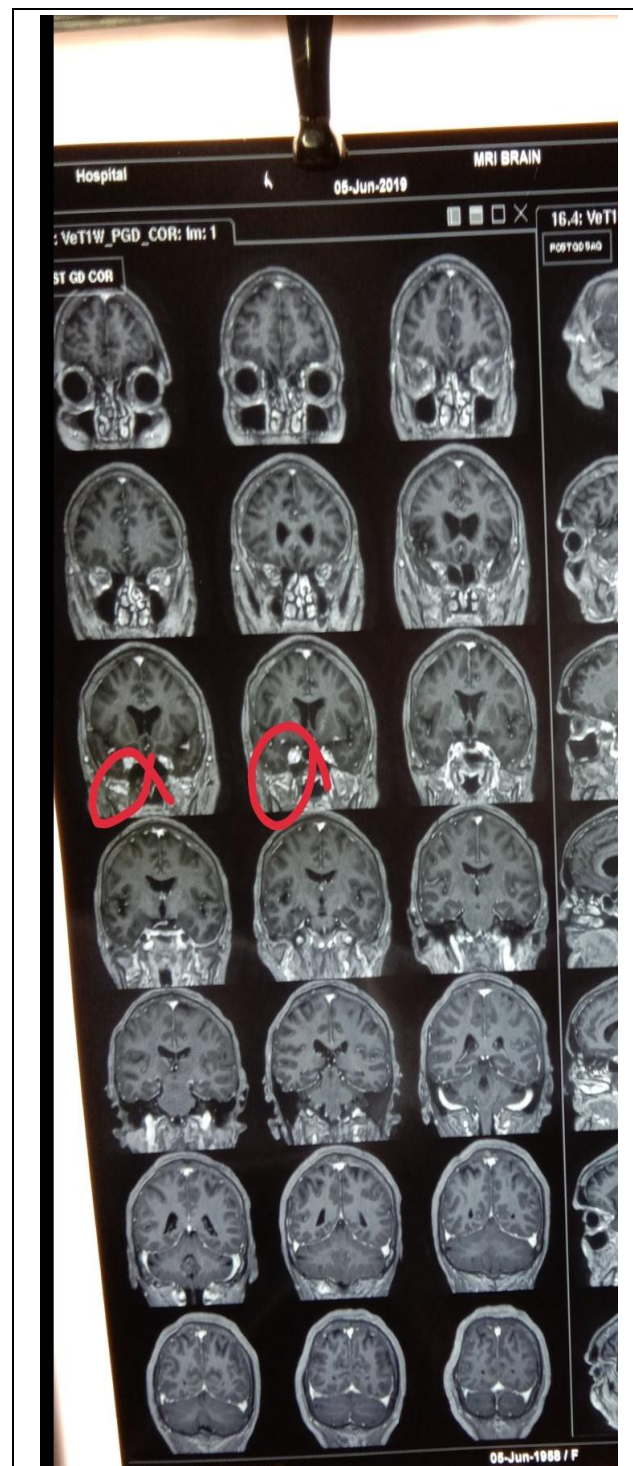


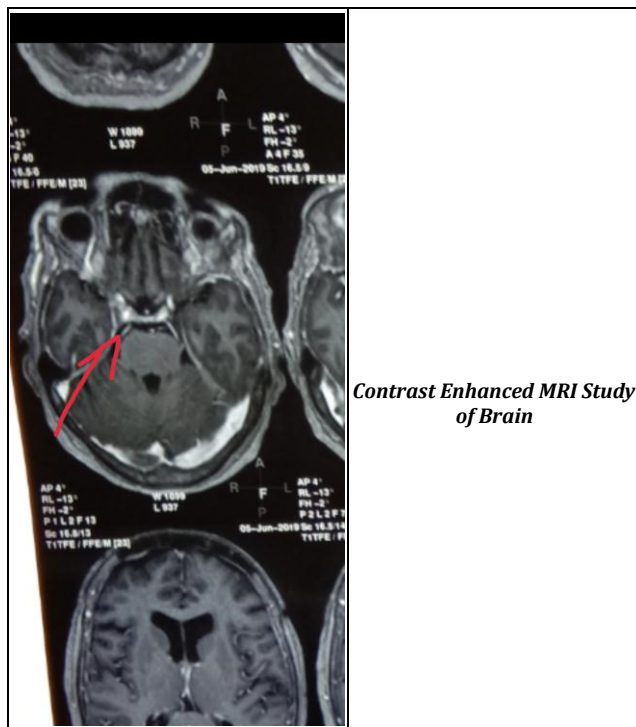
Figure 2. Presentation on Day 6 after Steroid Therapy

PATHOLOGICAL DISCUSSION

Her haemoglobin was 11.3 gm/dL, with total leucocyte count of 6800/cumm with differential count 62/12/2/1, normal



MRI Brain with Contrast was done, Abnormal Finding with Contrast- Contrast Enhancing Lesion Seen



Contrast Enhanced MRI Study of Brain

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 differential diagnosis, including meningioma, sarcoidosis, pituitary tumours, tuberculous meningitis (TBM) and lymphoma.^[8]

MRI findings classically demonstrate a soft-tissue mass lesion involving the SOF 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.^[9] Significant enhancement of the mass lesion is demonstrated on CE sequences. Of particular value is the post-contrast fat-saturated thin-slice coronal images through the orbital apex and cavernous sinus.

DISCUSSION OF MANAGEMENT

We started treatment with steroid. Response was very quick. Within 24 hours there was symptomatic relief in pain and within 6 days there was reverting of ptosis.

CONCLUSIONS

Tolosa hunt syndrome is a diagnosis of exclusion. Careful history taking, examination, positive neuroimaging finding, and steroid response are considered for diagnosis.

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