

Tolosa-Hunt Syndrome: a rare case report

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Abstract: 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 right orbital apex without any e/o filling defect in cavernous sinus or dilated SOV suggestive of THS. Other differential diagnosis were excluded on the basis of careful history, examination and investigations.

Keywords: cavernous sinus, Tolosa-Hunt syndrome, painful ophthalmoplegia, ptosis

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I. Introduction

Almost 64 years ago in 1954 Tolosa reported a patient with left orbital pain, ipsilateral progressive visual loss, total left ophthalmoplegia, and reduced sensation over the first division of the trigeminal nerve. Cerebral angiography disclosed narrowing of the intracavernous segment of the left internal carotid artery¹.

Seven years later, Hunt *et al* defined a clinical entity “of somewhat obscure aetiology” on the basis of six patients. In one patient surgical exploration of the parasellar region showed all structures to be “intact and healthy”. The authors thought that this syndrome of painful ophthalmoplegia was caused by an inflammatory lesion in the cavernous sinus, as described by Tolosa².

In 1966, Smith and Taxdal apply the eponym “Tolosa-Hunt syndrome”³.

Tolosa-hunt syndrome is a rare clinical entity with annual incidence 1 per million worldwide⁴ 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 steroid.

In most cases etiology is unknown. THS is thought to be caused by nonspecific granulomatous inflammation of cavernous sinus and superior orbital fissure that may extend into orbital apex. In the later situation vision may be diminished due to involvement of optic nerve.

II. Case Report

A 40 year old female came to the ophthalmology OPD with complaints of headache right side since one month and drooping of right upper eye lid for about the same duration. On elaborating history the headache was sudden in onset, throbbing in nature involving the right half and around the right eye, extending to frontal and temporal region and associated with nausea on and off. Three days after patient developed ptosis of right side. Patient was having similar complaints on the same side one year back for which she was admitted for 2 weeks and was managed conservatively. Patient was non diabetic and non-hypertensive. There was no history of head trauma.

On examination the patient was afebrile, with pulse rate 82 beats/min and blood pressure 130/80mmHg. There were no signs of pallor, icterus, cyanosis, clubbing, raised jugular venous pressure, and pedal edema. She was conscious and oriented and other higher mental functions were normal.

On ocular examination (Figure 1) there was complete ptosis of right side at the time of presentation. BCVA for right eye was counting finger close to face and left eye was 6/6, N6. Corneal sensation diminished on right side. Pupil was mid dilated non-reacting to light but 2 days after the admission (before the commencement of treatment) she developed grade 1 RAPD. Rest of the anterior segment was within normal limits.

There was complete ophthalmoplegia (Figure 2) on the right side. Examination of the left eye was normal. Fundus examination was normal in both eye.

There was reduced sensation over the right half of face above the angle of mouth suggestive of paralysis of first and second division of trigeminal nerve. Examination of other cranial nerve was normal. Other system examination was essentially normal.



Figure 1: Complete ptosis of right side.



Figure 2: Complete ophthalmoplegia of right side (right upper eye lid was lifted manually.)

On investigations, her hemoglobin was 11.2 gm/dL, with total leucocyte count of 6600/mm³. Blood sugar was 87 mg/dL with normal liver and renal function tests. Her ESR was 13 mm in first hour and serum C - reactive protein was 0.22mg/dl. Thyroid function test was within normal limits. Coagulation profile and serum lipid profile was also found to be normal. Mantoux test was negative. Human immunodeficiency virus testing by enzyme-linked immunosorbent assay was negative. ANA and ANCA testing was found to be negative. ECG, X-ray chest, and ultrasound abdomen revealed no abnormal findings. USG B-scan of right eye was normal.

Radiological investigation in the form of NECT brain with orbit was advised but revealed no abnormality. MRI brain was advised and findings were as below (Figure 3)

- T2 isointense extra axial soft tissue thickening noted in the right cavernous sinus extending anteriorly into the right orbital apex. On post contrast scan the homogenous enhancement leading to lateral bulging of right cavernous sinus without compromising right cavernous ICA lumen.
- No evidence of filling defect noted in the cavernous sinus. The maximum thickness of enhancing soft tissue in cavernous sinus measures 7.4mm.
- Impression: enhancing soft tissue thickening along right cavernous sinus anteriorly extends into the right orbital apex without any filling defect in the cavernous sinus or dilated SOV, suggestive of Tolosa-Hunt Syndrome.

Rest of the differentials of cavernous sinus involvement were ruled out on the basis of history, physical examination, and relevant investigations. The patient was started with intravenous dexamethasone (4mg/kg) for initial 3 days and oral prednisolone (1mg/kg) thereafter tapered weekly.

Patient responded well to the treatment. Headache subsided within 24 hours of treatment. After 2 days there was no RAPD but pupil reaction was sluggish. After 1 week ptosis improved, vision improved to 1/60 in right eye, pupil reaction was normal, slight eye movement in down gaze. After 2 weeks ptosis improved (Figure 4), vision improved to 2/60 in right eye, pupil reaction was normal, eye movement improved in all gazes. (Figure 4)

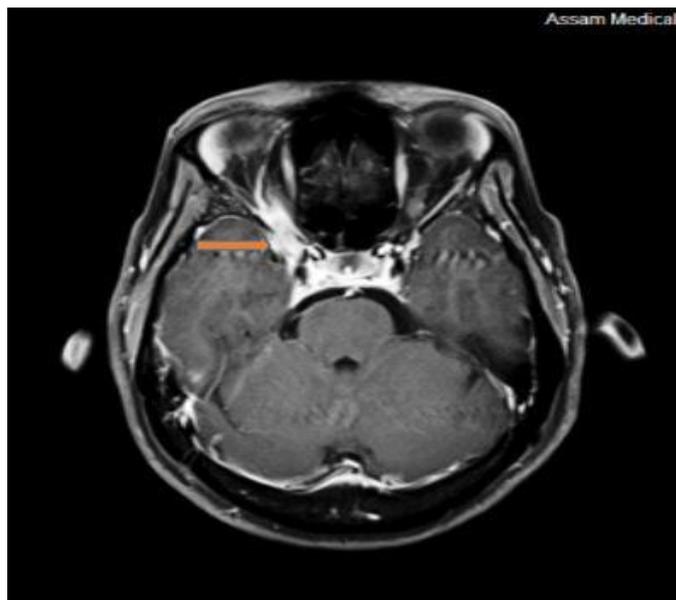


Figure 3: MRI Brain with T2 isointense extra axial section showing soft tissue thickening noted in the right cavernous sinus extending anteriorly into the right orbital apex.



Figure 4:Extra ocular movement of right eye after 2 week of treatment.

On the basis of history, typical clinical features on examination, findings on radiological investigations and excluding other possible conditions the diagnosis of Tolosa – Hunt Syndrome was made.

III. Discussion

Tolosa-Hunt Syndrome is 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). Pathologically, the inflammatory process is characterized by noncaseating, giant cell granuloma, fibroblast, lymphocyte, and plasma cell proliferation within the cavernous sinus septa and its walls^{1, 2}. The infiltration of the cavernous sinus with a non-specific inflammatory tissue leads to compressive neuropathy of the cranial nerves third, fourth, and sixth and V1 and V2 segments of trigeminal nerve, inside the cavernous sinus. The inflammatory process may also extend beyond the cavernous sinus/SOF, as is supported by involvement of other nerves such as optic and facial nerves in some case reports^{5, 6, 7, and 8}, as seen in our case.

Worldwide incidence 1 in 1 million annually. Only one case in New Zealand and New south wales till now³. 1st to the 8th decade (rare before 20 yrs.) with no sex predilection⁹.

The International Headache Society (IHS) criteria for THS^{13, 14} include the following:

- Episode(s) of unilateral orbital pain for an average of 8 weeks if left untreated.
- Associated paresis of the third, fourth, or sixth cranial nerves, which may coincide with onset of pain or follow it by a period of up to 2 weeks.
- Pain that is relieved within 72 h of steroid therapy initiation.
- Exclusion of other conditions by neuroimaging and (not compulsory) angiography.

Contrast enhanced MRI with multiple views, particularly coronal sections, should be the initial diagnostic study performed. The major limitation of MRI findings in Tolosa-Hunt syndrome is their lack of specificity. Yousem *et al* examined 11 patients and reported pathological MRI findings in the cavernous sinus in nine. In six of these nine the affected cavernous sinus was enlarged; in five of nine it had a convex lateral wall. Extension into the orbital apex was seen in eight patients¹⁰.

Resolution of pain within 24-48 hours after the initiation of therapy is characteristic¹¹. Resolution of imaging abnormalities after a course of systemic corticosteroids should be considered “diagnostic” of Tolosa-Hunt syndrome¹². Relapse is common within months to years in 30-40% of cases who have been successfully treated¹².

IV. Conclusion

THS is a diagnosis of exclusion. Each case of painful ophthalmoplegia should be investigated to rule out other common causes. CEMRI Brain and orbit with multiple view particularly coronal section is very helpful to identify the pathology of cavernous sinus in case of THS. Respond to steroid within 24-48 hours is characteristic. Relapse is quite common. Resolution of MRI findings after a course of steroid is considered diagnostic.

References

- [1]. Tolosa E. Periarteritic lesions of the carotid siphon with the clinical features of a carotid infraclinoid aneurysm. *J Neurol Neurosurg Psychiatry* 1954; 17:300–2.
- [2]. Hunt WE, Meagher JN, Le Fever HE, et al. Painful ophthalmoplegia. Its relation to indolent inflammation of the cavernous sinus. *Neurology* 1961; 11:56–62.
- [3]. Smith JL, Taxdal DSR. Painful ophthalmoplegia. The Tolosa-Hunt syndrome. *Am J Ophthalmol* 1966; 61:1466–72.
- [4]. G. Iaconetta, L. Stella, M. Esposito, and P. Cappabianca, “Tolosa-Hunt syndrome extending in the cerebello-pontine angle,” *Cephalalgia*, vol. 25, no. 9, pp. 746–750, 2005.
- [5]. Pirastehfar M, Luo JJ. Tolosa-Hunt syndrome with facial nerve paresis. *Neurological cases* 2014; 2:13–16.
- [6]. Mathew NT, Chandy J. Painful ophthalmoplegia. *J Neurol Sci* 1970; 11:243–56.
- [7]. Schatz NJ, Farmer P. Tolosa-Hunt syndrome: the pathology of painful ophthalmoplegia. In: *Neuroophthalmology Symposium of the University of Miami, and Boscom Palmer Eye Institute*, vol VI, Smith JL (Ed.). St Louis: Mosby, 1972. pp. 102–12.
- [8]. Spinnler H. Painful ophthalmoplegia; the Tolosa-Hunt syndrome. *Med J Aust* 1973; 2:645–6.
- [9]. Tessitore E, Tessitore A (2000) Tolosa-Hunt syndrome preceded by facial palsy. *Headache*40:393-396 & Basu A, MandalA (2014) "Tolosa Hunt Syndrome: Reported from West Bengal, India.
- [10]. Yousem DM, Atlas SW, Grossman RI, et al. MR imaging of Tolosa-Hunt syndrome. *AJNR Am J Neuroradiol* 1990; 10: 1181–4.
- [11]. Neigel JM, Rootman J, Robinson RJ, Durity FA, Nugent RA (1989) The Tolosa-Hunt syndrome: Computed tomographic changes and reversal after steroid therapy, *Can J Ophthalmol*21:287-290.
- [12]. Hannerz J (1999) Recurrent Tolosa-Hunt syndrome: a report of ten new cases, *Cephalalgia*19:33-35. (<http://onlinelibrary.wiley.com/doi/10.1111/j.1468-2982.1999.tb00055.x/abstract>).
- [13]. Colnaghi S, Versino M, Marchioni E, Pichiecchio A, Bastianello S, Cosi V, et al. ICHD-II diagnostic criteria for Tolosa-Hunt syndrome in idiopathic inflammatory syndromes of the orbit and/or the cavernous sinus. *Cephalalgia* 2008; 28(6):577–8.
- [14]. [Guideline] The International Classification of Headache Disorders, 2nd edn. *Cephalalgia* 2004; 1:9–160.

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