Case Report

A Rare Case of Superior Orbital Fissure Syndrome

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ABSTRACT

TOLOSA HUNT SYNDROME (THS) is a very rare entity with an annual incidence of one case per million per year. It is a diagnosis of exclusion and is characterised by painful ophthalmoplegia due to non specific inflammatory process in the region of cavernous sinus/superior orbital fissure. This 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, localising the pathological process to the region of the Cavernous sinus / Superior orbital fissure.

We report a interesting case of THS in a 38 year female who presented with unilateral headache associated with progressive diminution of vision and drooping of the ipsilateral eye lid. After diagnostic work up,we concluded that the patient had Tolosa Hunt Syndrome. Patient was initiated on steroids and responded well with significant improvement in 5 days and complete improvement on 15 days follow up.

Introduction :

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.[1]

The constellation of findings described may be due to four major causes : trauma, neoplasm, aneurysm, and inflammation

Tolosa originally described THS secondary to non-specific, chronic inflammation with proliferation of fibroblasts and infiltration of the septa and wall of the cavernous sinus with lymphocytes and plasma cells[2]. Hunt et al corroborated these findings, emphasizing the lack of necrosis and pointed out that "such inflammatory changes, in a tight connective tissue, may exert pressure upon the penetrating nerves."[3] Subsequent reports have shown granulomatous inflammation, with epithelioid cells and occasional giant cells.[5] Necrosis may also be seen. There have been no reports of an infectious organism associated with Tolosa-Hunt syndrome.

IHS (international Headache Society) laid down criteria for the diagnosis of THSin 1988,[6] which were modified in 2004[7].

A. Unilateral orbital or periorbital headache fulfilling criteria C
B. Both of the following :
   1. granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy
   2. paresis of one or more of the ipsilateral IIIrd, IVth and/or VIth cranial nerves
C. Evidence of causation demonstrated by both of the following :
   1. Headache is ipsilateral to the granulomatous inflammation
   2. Headache has preceded paresis of the IIIrd, IVth and/or VIth nerves by =2 weeks, or developed with it.
D. Not better accounted for by another ICHD-3 diagnosis

Treatment :

Almost 40 years ago Hunt first documented the beneficial effect of corticosteroid therapy in Tolosa-Hunt syndrome. Unfortunately, since then there is

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little new information as to optimal dosage, duration of treatment, or alternative forms of therapy. Treatment should be with high dose steroids (1 mg/kg/d) tapered slowly over 3 to 4 months[1]

Case Report :

This 38 year female, resident of Seoni, MP was admitted with complaints of left-sided headache associated with periorbital pain for 1 month along with gradual diminution of vision in the left eye for 15 days, followed by complete loss of vision for the last 15 days. She also complained of progressive drooping of the left eyelid for the initial 15 days with complete drooping since last 15 days. There was no history of vomiting, fever, aura, congestion of eyes, photophobia, joint pains, oral ulcers, rash, limb weakness, any unknown bite, waxing and waning of complaints. She was not a known case of DM, HTN, Thyroid disorder, Koch's. Patient's general examination was within normal limits.

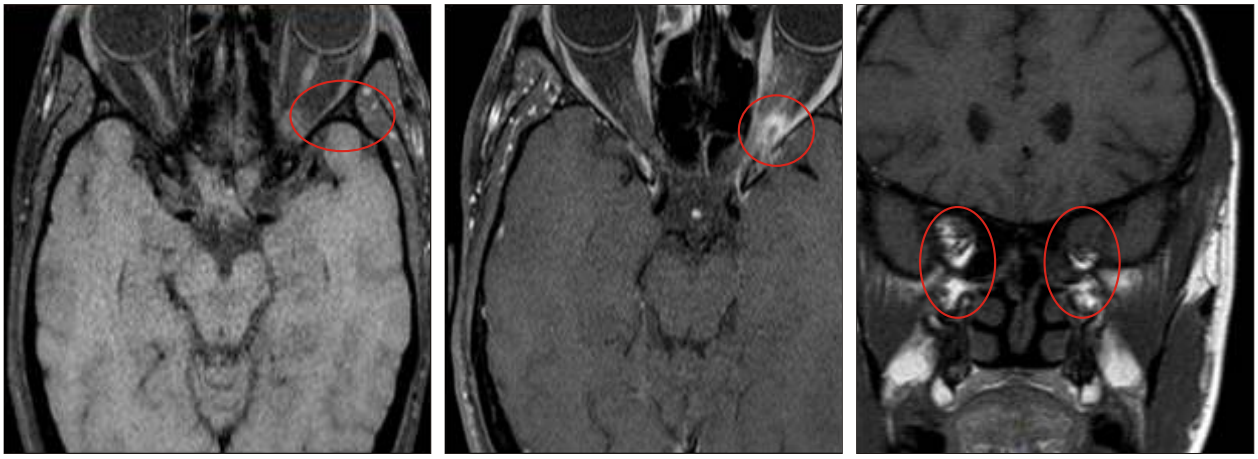
On CNS examination higher mental functions were normal.

- On Cranial nerve examination, optic nerve examination revealed absent perception of light in the left eye, right eye was normal and bilateral fundus examination also was WNL.
- On 3rd, 4th, 6th cranial nerve examination; left complete ptosis was present along with dilated pupil and absent direct and consensual light reflex and accommodation reflex. In the Right eye, pupil was normal-sized along with preserved direct light reflex and accommodation reflex, although consensual light reflex was absent. On ocular movement examination, complete ophthalmoplegia was present in the left eye with normal movements in the right eye.
- On 5th cranial nerve examination, the Motor examination was normal B/L with hypoaesthesia in left ophthalmic division. Corneal reflex was diminished on the left side with normal jaw jerk.
- Rest all cranial nerve examination were WNL. Motor and Sensory system examination was WNL. All superficial and deep tendon reflexes were WNL. No involuntary movements were present. No Cerebellar signs were seen. Gait was normal.
- The provisional diagnosis was kept as a case of Left-Sided Multiple Cranial Nerve Palsy (2ND/3RD/4TH/5TH (V1)/6TH)
- Possibility of Cavernous Venous Sinus Thrombosis / Superior Orbital Fissure Syndrome / Autoimmune etiology / ICSOL was kept.
- Battery of investigations involving CBC, LFT, KFT, Serum Electrolytes, Lipid Profile, ANA, HIV, HBsAg, VDRL, ESR, Thyroid Profile, ANCA, ACE Levels were done. All of them were inconclusive.
- CECT HEAD + CT ORBIT : was done which was showed thickened left optic nerve sheath possibly? optic neuritis.
- MRI was done and was suggestive of an Ill-defined hypointensity of approx size 8x5 mm in the intraconal compartment of the left eye with extension along the left orbital apex in close proximity with the canalicular segment of optic nerve compressing and displacing it medially with its segmental thinning. A T2/STIR hyperintensity noted in the left extraocular muscles which appeared bulky and showed heterogeneous post-contrast enhancement with no obvious pre septal edema. No abnormal signal intensity in Optic Nerve and Bilateral cavernous sinuses showed normal opacification. Features were suggestive of **LEFT ORBITAL APEX SYNDROME.**

The final diagnosis was kept as c/o left-sided multiple cranial palsy secondary to superior orbital fissure syndrome? Cause

Since the studies showed no abnormalities and we excluded neoplastic, infectious, vascular, thyroid and metabolic causes of painful ophthalmoplegia, we started treatment for Tolosa-Hunt Syndrome with Methylprednisolone 1 gram IV daily for 5 days

Patient started showing improvement after the 3rd dose of methylprednisolone with the gradual improvement of ptosis and perception of light. External ophthalmoplegia started improving after



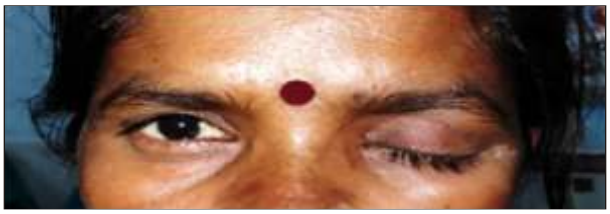
3rd dose and improved completely except for lateral rectus palsy. The patient was discharged thereafter followed by Tab. Prednisolone in tapering doses. On 1 month follow up patient recovered fully without any neurological deficit.

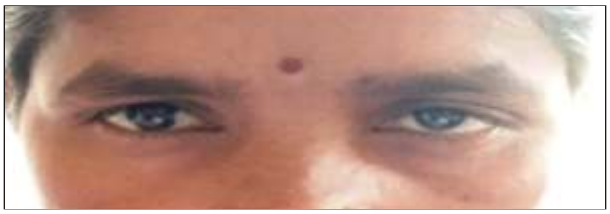
As no specific cause could be found out after detailed evaluation as well as the steroid responsiveness of the patient's clinical condition, the diagnosis was kept as TOLOSA HUNT SYNDROME.

LEFT EYE BEFORE TREATMENT

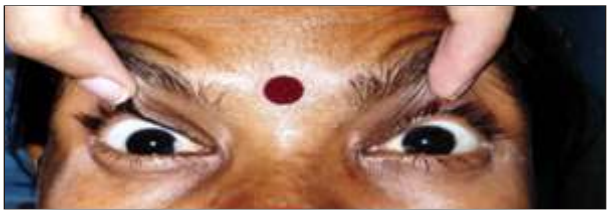
LEFT EYE AFTER TREATMENT

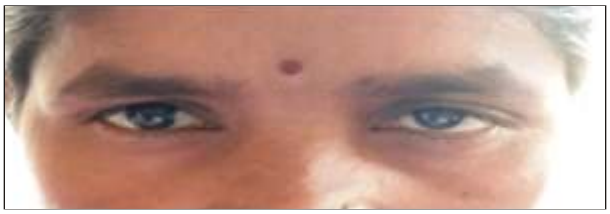
Ptosis



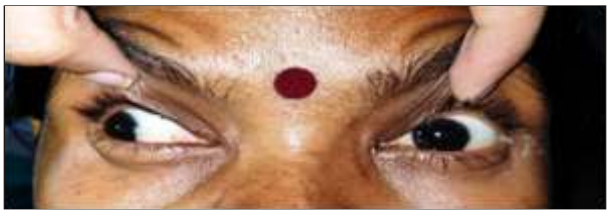


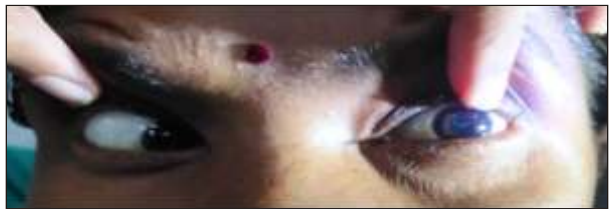
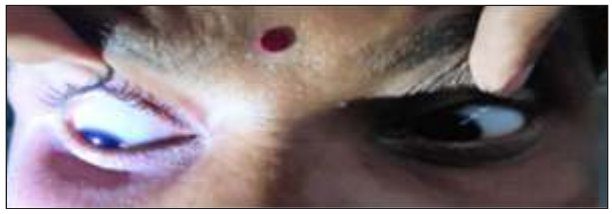
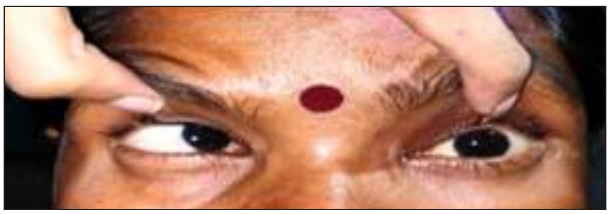
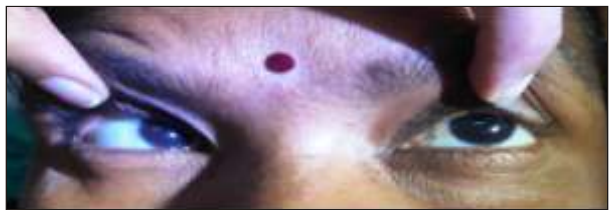
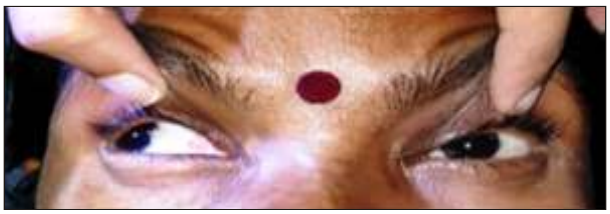
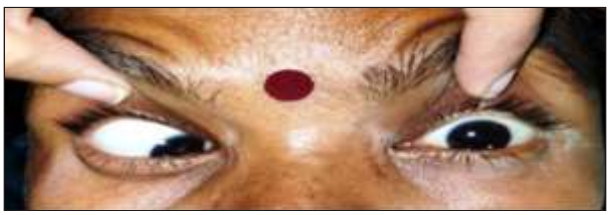
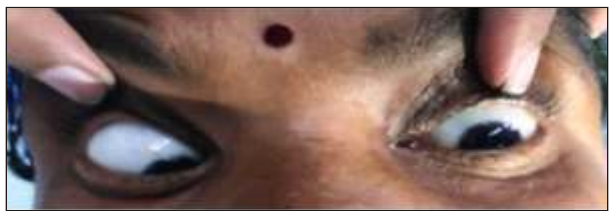
Primary Gaze





Medial Rectus



LEFT EYE BEFORE TREATMENT	LEFT EYE AFTER TREATMENT
Lateral Rectus	
	
Superior Oblique	
	
Superior Rectus	
	
Inferior Oblique	
	
Inferior Rectus	
	

Discussion :

Tolosa-hunt syndrome is a rare syndrome with an estimated annual incidence of one case per million per year and is thought to be caused by idiopathic granulomatous inflammation of the cavernous sinus. THS is known to be exquisitely responsiveness to glucocorticoid treatment. Though considered a benign condition, permanent

neurologic deficits can occur, and relapses are common, often requiring prolonged immuno suppressive therapy.

The typical presentation of headache followed by ophthalmoplegia was present in our patient. As mentioned in IHS guidelines, there was also the involvement of the ophthalmic branch of the trigeminal nerve in the form of hypoaesthesia in the distribution of respective nerve on top of the classic oculomotor, trochlear, and abducens cranial nerve palsies.

Most patients (greater than 75 percent) who present with painful ophthalmoplegia will not have Tolosa-hunt syndrome. The syndrome of painful ophthalmoplegia may be caused by any process exerting a mass effect on the cavernous sinus. These include a primary intracranial tumor, lymphoma or other local or distant metastatic tumors, aneurysm, carotid-cavernous fistula, carotid dissection, cavernous sinus thrombosis, infection, vasculitis, and sarcoidosis. Of these conditions, tumors and vascular conditions are the most common. As suggested by MRI, all these conditions were ruled out in our patient.

Cases of Tolosa-hunt syndrome have also been reported in patients with inflammatory disorders, such as systemic lupus erythematosus, but this may simply represent an association of the two autoimmune conditions. Howsoever, SLE was ruled out in our patient as her ANA was negative.

Cases of orbital inflammation may be the initial presentation of systemic inflammatory disorders such as sarcoidosis and Wegener's granulomatosis which were ruled out in our case as ACE levels and ANCA were not affirmative.

In addition to these structural compressive lesions, painful ophthalmoplegia can also be caused by diabetic cranial nerve palsy, but HbA1c of our patient came out to be in non-diabetic range, as well in-hospital monitoring of blood sugar levels ruled out diabetes as a cause for her condition. Moreover, diabetes more commonly causes mononeuropathy rather than polyneuropathy.

Orbital pseudotumor is a related condition of idiopathic inflammation involving the orbit. Patients present with painful ophthalmoplegia and orbital signs (proptosis, conjunctival injection, and chemosis). This condition, which is also glucocorticoid responsive, may be identical to Tolosa-hunt syndrome, distinguished only by a different anatomic localization. Therefore, distinguishing between this syndrome and Tolosa-hunt syndrome is less important than excluding other causes of painful ophthalmoplegia, but as the MRI was suggestive of the involvement of superior orbital fissure only and no evidence of any intraorbital enhancement or mass was present, this condition was also less likely in our patient.

Our patient met the aforementioned criteria and all space-occupying,vascular and autoimmune etiologies were ruled out making a diagnosis of THS more likely

Another important consideration while managing THS is relapses which occur in about one-half of reported patients over an interval of months to years, and appear to be more likely in patients who are younger. Ipsilateral, contralateral, and bilateral relapses have been reported. hence a strict follow up is necessary for years. Managing relapses require repeated investigations to rule out inflammatory and neoplastic disorders such as Sarcoid, Wegener's granulomatosis, and lymphoma.

Conclusion :

In general, it is difficult to exclude alternative diagnoses by clinical features alone. Neuroimaging and other diagnostic tests are generally required. Other clinical signs may be suggestive but are not completely reliable. This case report demonstrates the importance of including THS as a differential diagnosis after all other diagnostic studies rule out more common etiologies. Patient should also be followed up meticulously for early recognition of relapses.

References :

1. Gladstone JP. An approach to the patient with painful ophthalmoplegia, with a focus on Tolosa-Hunt syndrome. Curr Pain Headache Rep. 2007 Aug;11(4):317-25.
2. Tolosa E. Periarteritic Lesions of the Carotid Siphon with the Clinical Features of a Carotid Infraclinoidal Aneurysm. J Neurol Neurosurg Psychiatry. 1954 Nov;17(4):300-2.
3. Hunt WE, Meagher JN, Lefever HE, Zeman W. Painful opthalmoplegia. Its relation to indolent inflammation of the carvernous sinus. Neurology. 1961 Jan;11:56-62.
4. Smith JL, Taxdal DS. Painful ophthalmoplegia. The Tolosa-Hunt syndrome. Am J Ophthalmol. 1966 Jun;61(6):1466-72.
5. Schatz N, Farmer P. Tolosa-Hunt syndrome: the pathology of painful ophthalmoplegia. In 1972. p. 102-12.
6. Headache Classification Committee of the International Headache Society. Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. Cephalalgia. 1988;8(7):1-96.
7. Lance JW, Olesen J: The International Classification of Headache Disorders ICHD-II. Cephalalgia 2004, 24:131.
8. https://eyewiki.aao.org/Tolosa-Hunt_syndrome