RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 5 | May 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

Rare Case of Ophthalmoparesis (Tolosa Hunt Syndrome)

KEYWORDS: Tolosa Hunt syndrome, ophthalmoparesis

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ABSTRACT *TolosaHunt syndrome(THS) is a painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure.*

1. INTRODUCTION

THS is caused by an inflammatory process first described by Tolosa(1) as granulomatous periarteritis of cavernous carotid, Hunt et al(2) described proliferation of fibroblasts and infiltration of septa and wall of cavernous sinus with lymphocytes and plasma cells. Its cause is unknown. Pathologically this syndrome falls within the range of idiopathic orbital pseudotumor(3)

CASE STUDY:

- 18 year old female patient came with complaints of double vision, painful movements of left eye,paraesthesias of left side of forehead since 2 months.
- No history of redness of eye ,fever, weight loss, head ache, loss of vision.
- Past history: History of cervical tuberculous lymphadenitis 2 years back for which she completed treatment.
- Family history: Not significant
- Personal history: mixed diet ,normal appetite, bowel and bladder regular.
- Birth history: Full term normal vaginal delivery, institutional delivery, born out of non consanguineous marriage, cried immediately after birth.
- GENERAL EXAMINATION: Patient conscious, coherent, no pallor ,cyanosis, clubbing, edema, lymphadenopathy
- Vitals : stable
- CVS,GIT,RESPIRATORY SYSTEM EXAMINATION: Normal
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- CNS EXAMINATION:
- Partial restriction of left lateral rectus, left inferior oblique muscle movement(inability to adduct and elevate),as shown in fig 1, mildly dilated and sluggishly reacting pupil on the left, decreased sensation on the forehead i.e in the first division of left trigeminal nerve, absent corneal reflex on the left. Rest of the nervous system examination unremarkable.
- INVESTIGATIONS:
- Hb:11 gms, WBC:6400, Platelets: adequate
- ESR: 12 mm first hour
- CUE: Normal
- RFT, LFT, Electrolytes: Within normal range
- Serum calcium: Normal
- HIV, HBsAg: Non reactive
- CXR: Normal
- CSF Analysis: cell count 6 cells,94% lymphocytes, proteins, sugars- within normal range, ADA:8, bacterial gram stain, culture/sensitivity, fungal culture: sterile, TB PCR: Negative
- CT Brain and both Orbits: Normal
- MRI Brain and both Orbits(plain and contrast): ill defined granulomatous lesion involving left cavernous sinus and adjacent dura extending upto orbital apex suggestive of THS as shown in fig 2.
- MRV Brain: Normal
- Fundus examination: Normal

DIAGNOSIS: LEFT SIDED MULTIPLE CRANIAL NERVE PALSIES(3,5,6) CAUSED BY TOLOSA HUNT SYNDROME

Partial paralysis of left inferior oblique muscle and left lateral rectus muscle

Fig 1

fig:2

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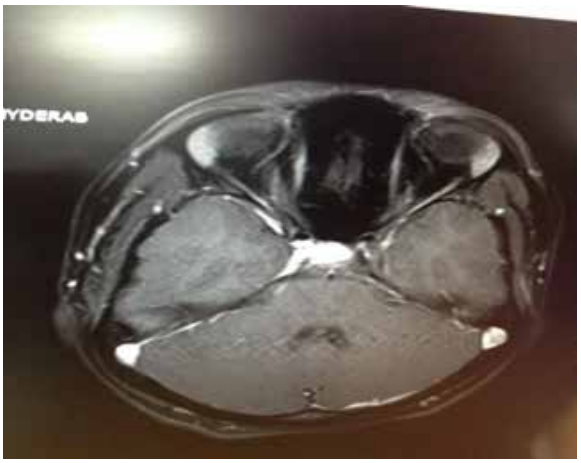

Fig 2
Ill defined granulomatous lesion involving left cavernous sinus and adjacent dura exteing upto left orbital apex

2.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 seen(4).

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 cause of the syndrome. Pathologically, infiltration of lymphocytes and plasma cells as well as thickening of the dura mater was seen(4). The condition was termed Tolosa-Hunt syndrome by Smith and Taxdal in 1966(5).The latter authors stressed the importance of the dramatic rapid response to steroid therapy.

In 1988 THS criteria were provided by the International Headache Society and further revised in 2004

INTERNATIONAL HEADACHE SOCIETY CRITERIA FOR THS:(6,7)

- Episode of unilateral orbital pain for an average of 8 weeks if left untreated.
- Associated paresis of 3,4,5,6 cranial nerves which may coincide with onset of pain or follow it by a period of upto 2 weeks.
- Pain that is relieved within 72 hours of steroid therapy initiation.
- Exclusion of other conditions by neuroimaging and angiography(not compulsory).

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(5).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(7).

Our patient responded to steroids with a decrease in pain, ophthalmoparesis showing partial improvement.

CONCLUSION: Administration of systemic steroids for 48 hours in a patient with THS produces a dramatic response in painful ophthalmoplegia that allows differentiation of this cause from other conditions of painful ophthalmoplegia.(8)

REFERENCES:

1, Tolosa EJ: Periarteritic lesions of the carotid siphon with clinical features of carotid intraclinoid aneurysmus. J Neurol Neurosurgery Psychiatry 1954; 17:300-302.

2, Hunt WE, LeFever H: Painful ophthalmoplegia: its relation to indolent inflammation of the cavernous sinus.Neurology 1961; 11:56-62.

3, Levy IS, Wright JE:Orbital & retro-orbital pseudotumour. Modern problems of ophthalmology 1975; 14:364-67.

4, Som PD,Curtin HD. Head and Neck Imaging. 4th ed. St LOUIS: Mosby, 2003:587-591.

5, Sathyanathan BP, Rajasundaram R, Sankaravadivelu ST, Nadhamuni K. A case of Tolosa-Hunt Syndrome – MR imaging appearance. Ind J Radiol Imag 2006;16(1):97-98.

6, Mendez JA, Arias CR, Sanchez D, et al. Painful ophthalomoplegia of the left eye in a 19-year-old female, with an emphasis on Tolosa-Hunt syndrome: a case report. Cases Journal 2009;2:8271.

7, Mora-de-Onate J, Pascual-Perez-Alfaro R, Izquierdo- Vazquez C, Gonzalez-Ruiz M, Aguirrebena-Olmos A, Diez-Villalba R. Painful ophthalmoplegia (pseudotumour of the orbit and Tolosa-Hunt Syndrome). Arch Soc Esp Oftalmol 2007;82:509-512.

8, Yousem DM, Atlas SW, Grossman RI, Sergott RC, Savino PJ, Bosley TM. MR imaging of Tolosa-Hunt Syndrome. AJNR 1990;154:167-170.