 DOI: 10.18231/2455-8478 .2017.0024

Tolosa Hunt Syndrome – Painful ophthalmoplegia in a young female

Saroj Gupta[1,*], Mary Jenifa Tigga[2], Sagarika Laad[3], Pranav Saluja[4]

[1]Professor & HOD, Consultant, Siddharth Eye Hospital, Bhopal, Madhya Pradesh, [2,3,4]PG Student, Dept. of Ophthalmology, People's College of Medical Sciences & Research Centre, Bhopal, Madhya Pradesh

***Corresponding Author:**
Email: sarojini94@yahoo.co.in

Abstract

Tolosa-Hunt syndrome (THS) is recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or superior orbital fissure (SOF) and apex of the orbit. A young female presented to eye OPD with chief complains of unilateral headache with pain, swelling and drooping of left eye. MRI brain and orbit revealed homogenous enhancing soft tissue mass at the left orbital apex and cavernous sinus. Patient was given systemic corticosteroid in tapering dose. Follow up after one week of therapy showed improvement in ptosis and ocular movement.

Keywords: Cavernous sinus, Orbital apex, Ophthalmoplegia, Ptosis, Superior orbital fissure

Introduction

Tolosa-Hunt syndrome (THS) is recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or superior orbital fissure (SOF) and apex of the orbit. It is characterized by periorbital pain, headaches on the same side and diplopia. Typical clinical features are retro-orbital pain, with ptosis and restriction of the ocular movements due to involvement of the 3rd, 4th and 6th cranial nerve in THS.[1-3] These disease is recurrent and well responds to steroid.[4] The key diagnostic procedure is MRI. Findings on MRI before and after systemic corticosteroid treatment are important diagnostic criteria to make the final diagnosis of TSH and to differentiate it from other cavernous sinus lesion.[5]

Case Report

A 22 year old female came to OPD with complains of unilateral headache associated with mild pain in left eye with drooping of eyelid, swelling, as well as decrease in visual acuity since 8 days but there was no history of redness or trauma.

Her vitals were normal. On ocular examination visual acuity was 6/6 in right eye and 6/9 in left eye. She had severe ptosis, marginal reflex distance (MRD) was zero, upper lid crease absent, levator palpebrae superioris action was < 4mm in left eye (Fig. 1). These signs further deteriorated. On second day all the ocular movement was restricted except abduction, pupil was mid dilating (6mm) and direct pupillary reaction was sluggish. There was no facial sensory or motor loss in the distribution of the trigeminal nerve was detected. Funduscopy was normal. Complete blood count, blood sugar and routine urine were within normal limit. MRI brain and orbit shows lobulated focal lesion with partially ill defined margin at the region of anterior aspect left cavernous sinus and left orbital apex. The lesion measures **2.7× 1.2× 1.1cm** and shows isointense signal on T1W images and heterogeneous mixed signal in T2W images. On post contrast study lesion shows intense homogenous enhancement. The cavernous part left ICA was medially compressed and displaced medially by the lesion. The Meckel's cave was not involved. Both eye balls were normal in size, shape and position. It also reveals homogenous enhancing soft tissue mass at the left orbital apex and cavernous sinus (Fig. 3)

Patient was given systemic corticosteroid on 3rd day (Tab. Prednisolone 40mg) in tapering dose for 6week. On follow up after one week of therapy ptosis and ocular movement showed improvement (Fig. 2).

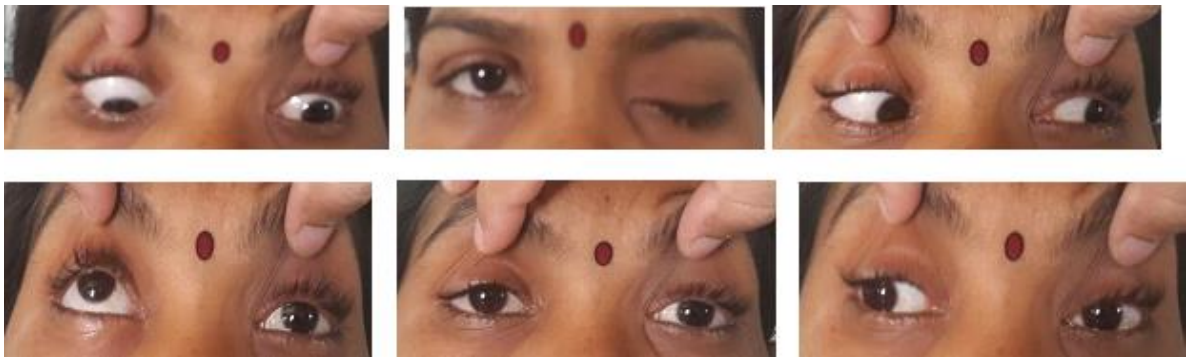

Fig. 1: Before Treatment

Fig. 2: After Treatment

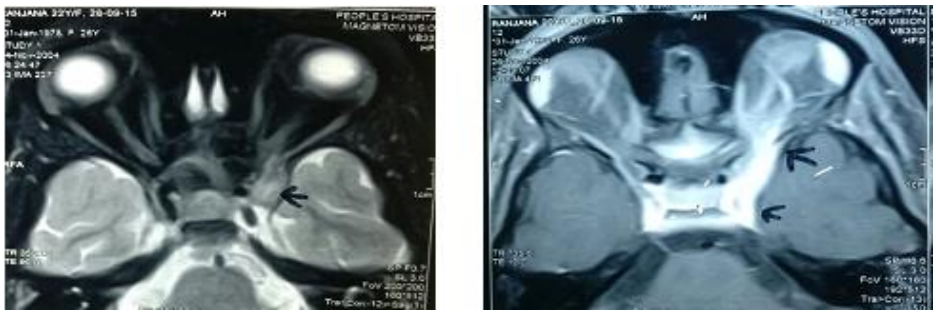

Fig. 3: Axial MRI image shows intense enhancement of the soft-tissue mass within the left SOF. The lesion extends into the cavernous sinus

Discussion

In 1954, Tolosa first described the condition with unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, VI and V. The patient was imaged using carotid angiography, and segmental narrowing of the carotid siphon was seen.[6] In 1961, Hunt et al. described similar clinical findings in 6 patients, 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.[6] Finally these condition was termed Tolosa-Hunt syndrome by Smith and Taxdal in 1966.[7] In 1988, THS criteria were provided by the International Headache Society (IHS), and further revised in 2004.[8,9] The cause of THS is still unknown. The pathological process is self-limiting and spontaneous remissions occur, a short course of steroids can dramatically relieve the periorbital pain within 24-48 hr, followed by gradual resolution of the cranial nerve dysfunctions.

MRI is the most sensitive investigating tool for THS. The differential diagnosis of THS including pituitary tumours, tuberculous meningitis (TBM), meningioma, sarcoidosis and lymphoma.[7] MRI findings are classically demonstrate a soft-tissue lesion involving the Superior Orbital Fissure or Cavernous Sinus. The characteristic features of THS on MRI is hypointense to fat and isointense to muscle on short TR/TE (repetition time/ eco time) sequences and isointense to fat on long TR/TE sequences.[10] On contrast MRI there is enhancement of lesion is characteristic features of THS.

Conclusion

THS patients present with painful Ophthalmoplegia. MRI is the most sensitive investigating tool for THS. The response to systemic corticosteroid is very good. Detail investigations including MRI contrast should be done to rule out other causes of painful Ophthalmoplegia.

References

1. Arar ZV, Janjetoviý Z, Mariniý M, Sekelj S, Lezaiý Z, Dikanoviý M. Painful ophthalmoplegia-Tolosa-Hunt syndrome. Acta Med Croatica 2007;61(4):395-8.
2. Gladstone JP. An approach to the patient with painful ophthalmoplegia, with a focus on Tolosa-Hunt syndrome. Curr Pain Headache Rep 2007;11(4):317-25.
3. Van Dalen JT, Bleeker GM. The Tolosa-Hunt syndrome. Doc Ophthalmol 1977;44(1):167-72.
4. Cakirer S. MRI findings in Tolosa Hunt syndrome before and after systemic corticosteroid therapy. Eur J Radiol 2003;45(2):83-90.
5. Cakirer S. MRI findings in the patients with the presumptive clinical diagnosis of Tolosa-Hunt syndrome. Eur Radiol 2003;13(1):17-28.
6. Som PD, Curtin HD. Head and Neck Imaging. 4th ed. St Louis: Mosby, 2003:587-591.
7. 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.
8. 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.
9. 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.
10. Yousem DM, Atlas SW, Grossman RI, Sergott RC, Savino PJ, Bosley TM. MR imaging of Tolosa-Hunt Syndrome. AJNR 1990;154:167-170.