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Indian Journal of Clinical and Experimental Ophthalmology

Journal homepage: www.ijceo.org

Case Report

A rare case of unilateral total ophthalmoplegia- Tolosa hunt syndrome

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Abstract

Tolosa hunt syndrome (THS) is a rare condition characterised by reversible painful ophthalmoplegia, episodic unilateral periorbital headache & paralysis of the 3rd, 4th &/or 6th cranial nerves which shows prompt response to steroids but can relapse & remit. Our patient is 23 years old female who presented with left eye painful total ophthalmoplegia with ipsilateral periorbital headache along with left eye ptosis & mild proptosis. Her all-routine investigations were within normal limits. MRI findings showed abnormality in left orbital apex and anterior aspect of cavernous sinus. After ruling out all differentials, the diagnosis of exclusion-Tolosa Hunt Syndrome was suspected. Patient when given trial of oral steroids showed drastic improvement. During follow-up visits, the drooping of the eyelid was decreased, and there was improvement in eye movements in all directions without pain or double vision. Early diagnosis not only decreases the severity & duration of disease but also reduces patient morbidity.

Keywords: Tolosa hunt syndrome, Painful total ophthalmoplegia, Ipsilateral headache, Cavernous sinus, Diagnosis of exclusion, Steroids.

Received: 04-08-2024; **Accepted:** 17-09-2024; **Available Online**: 09-06-2025

1. Introduction

Tolosa-hunt syndrome (THS) is named after Eduardo Tolosa and William Hunt, who independently identified cases of ophthalmoplegia accompanied by granulomatous inflammation in the cavernous sinus. The designation "Tolosa-Hunt Syndrome" was subsequently used for instances of cavernous sinus syndrome (CSS) where no specific cause could be pinpointed through diagnostic investigations. Despite its rarity, with an estimated annual incidence of 1 to 2 cases per million individuals, THS represents approximately 23% of all cavernous sinus syndrome cases. There is no known geographical, racial or gender preference associated with THS.[1,7] Tolosa Hunt Syndrome was first identified by the International Headache Society in 2004. According to the diagnostic criteria set forth in the International Classification of Headache Disorders, 3rd edition (ICHD-3), this condition is characterized by a unilateral headache located in the orbital or periorbital region. It is associated with granulomatous inflammation in the cavernous sinus or superior orbital fissure, which can be detected via MRI or biopsy. The syndrome also involves paresis of one or more of the ipsilateral cranial nerves—specifically the third, fourth, or sixth cranial nerves. The diagnosis is supported by the observation that the headache generally precedes the onset of cranial nerve weakness by less than two weeks, or occurs concurrently with it, and is located on the same side as the granulomatous inflammation.[2,6]

2. Case Report

A twenty-three year old female presented to ophthalmology OPD with the complaints of drooping of left eye upper eyelid and inability to move the left eyeball in any gaze since last 4-5 days. (**Figure 1**) It was associated with ipsilateral periorbital headache, diplopia, nausea and vomiting. There was no significant history of any ocular trauma, or any surgical intervention done recently. Patient was well built and adequately nourished without any systemic disease. On inspection patient had chin elevation without head tilt. The left eyebrow was at a higher level than the normal. Left eyeball was slightly proptosed. Left eye upper eyelid had severe ptosis with reduced Marginal Reflex Distance (MRD)-

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https://doi.org/10.18231/j.ijceo.2025.065

MRD1 of 1mm, MRD2 of 3mm and the palpebral fissure height of 4mm. On levator function test of the left eye the excursion was <4mm. Left eye pupillary reaction was sluggishly reactive. There were no signs of RAPD or anisocoria. Right eye anterior segment findings were within normal limits. Visual acuity (unaided) in both eyes was 6/6 when recorded on Snellen's chart. Extraocular movement examination revealed no movements in any gazes in left eye. (**Figure 2**) Movements were present in all gazes in right eye. Both eyes fundus examination was within normal limits. Her left eye ultrasonography (B-scan) showed no abnormality in posterior segment. Intraocular pressure in both eyes was 18 mm Hg on Perkins Tonometer. On Gonioscopic examination the angle was wide-open (Shaffer's System Grade-4). Her infectious & autoimmune work-ups were grossly negative. CSF examination was within normal limits. MRI findings showed T2 hyperintense signal of left orbital apex and anterior aspect of cavernous sinus. Both optic nerves displayed normal course, caliber and signal characteristics. (**Figure 3**) After ruling out all differentials, the diagnosis of exclusion-Tolosa Hunt Syndrome was suspected. Patient was given trial of oral steroids- she was started on Tab. Prednisolone 50mg OD for seven days, followed by tapering doses each week i.e. 40mg, 30mg, 20mg, 10mg, 5mg OD for seven days each. Patient responded drastically to steroid treatment. On subsequent follow-ups ptosis was reduced with improvement in extraocular movements in all gazes without pain or diplopia.

Figure 1: Initial presentation

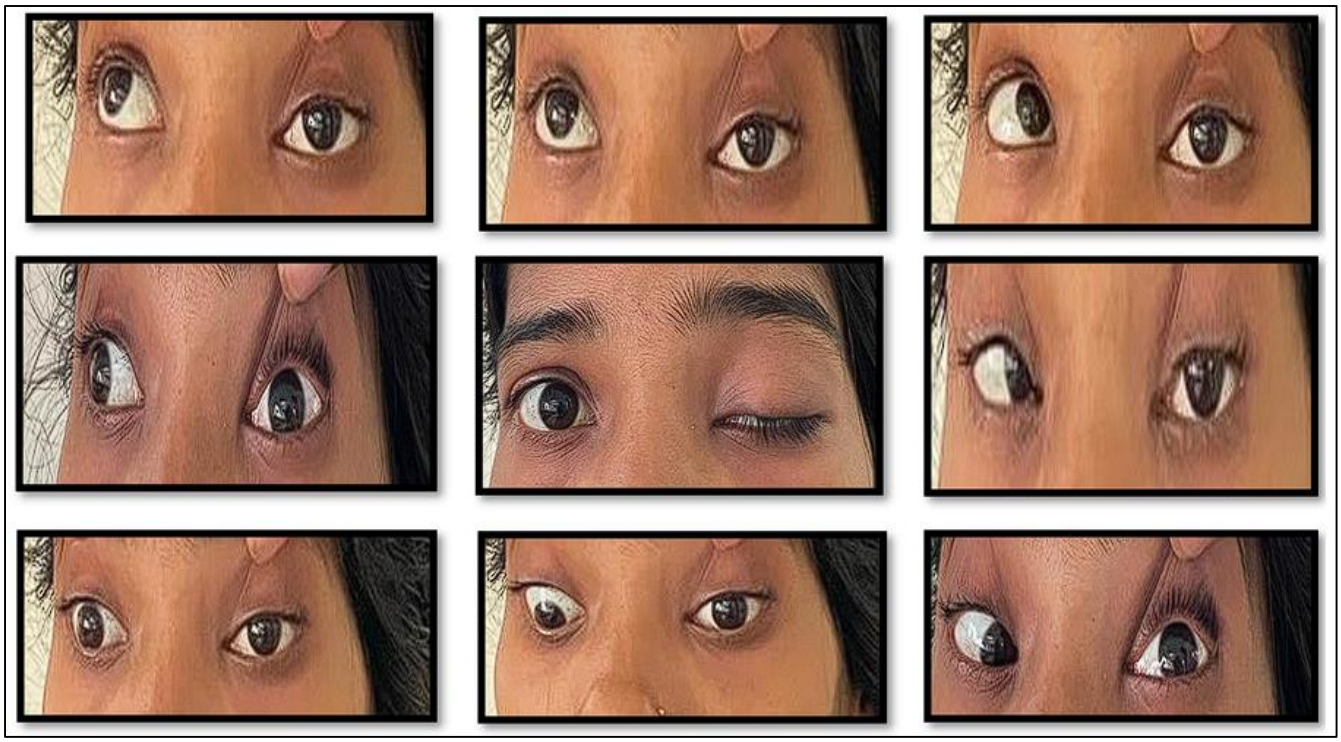

Figure 2: Left upperlid ptosis with restricted eye movements in all gazes

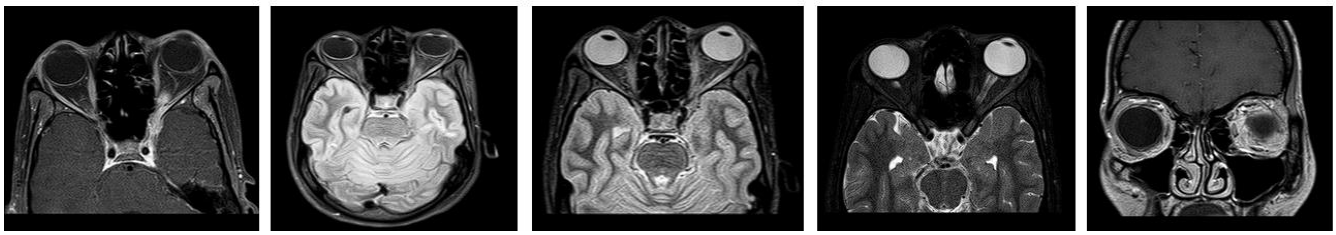

Figure 3: MRI findings: Hyperintense signals in left orbital apex and anterior aspect of cavernous sinus

3. Discussion

The exact cause of Tolosa-Hunt Syndrome (THS) remains unclear. However, biopsies and surgical findings have revealed granulomatous tissue containing fibroblasts and immune cells within the cavernous sinus. This tissue may lead to increased pressure on the nerves, potentially causing the observed paralysis.[3] MRI is the primary imaging technique used for diagnosis, as it offers more detailed visualization of the orbital apex and cavernous sinus compared to CT scans.[4] Oral Steroids are the mainstay of treatment. A key characteristic of THS that sets it apart from other causes of painful ophthalmoplegia is its swift and significant improvement in response to systemic steroids, often within just 48 hours.[5] Although the overall prognosis for THS is generally very good, the condition may follow a relapsing-remitting pattern.[8]

4. Conclusion

Tolosa-Hunt Syndrome is a diagnosis of exclusion that necessitates advanced imaging and laboratory tests for confirmation. Early and precise diagnosis through these methods allows for prompt treatment, which can lessen both the duration and severity of symptoms, including the risk of vision loss.

5. Source of Funding

None.

6. Conflict of Interest

None.

7. Acknowledgment

We would like to thank the patient for her cooperation and consenting for the publication of the photographs.

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Cite this article: Math SS, Gondhali GR, Rahul K. A rare case of unilateral total ophthalmoplegia- Tolosa hunt syndrome. *Indian J Clin Exp Ophthalmol*. 2025;11(2):365–367.