International Journal of Research and Review
[Indexed, Peer Reviewed Journal]
Volume 12; Issue: 1; January 2025
Website: www.ijrrjournal.com
Case Report *E-ISSN: 2349-9788; P-ISSN: 2454-2237*

Case Report on Tolosa Hunt Syndrome in a Diabetic Patient: An Uncommon Presentation

Ajay R[1], Shubham B Harapanahalli[1], Dr Veeranna Mohan Gadad[2], Gunupati Balamani Chandana[1], Gladys K Siji[1]

[1]Pharm D Interns, Department of Pharmacy Practice, Bapuji College of Pharmacy, Davanagere, Karnataka, India-577004,
[2]Associate Professor, Department of Neurology, SSIMS and RC, Davanagere, India-577004.

Corresponding Author: Shubham B Harapanahalli

DOI: https://doi.org/10.52403/ijrr.20250132

ABSTRACT

Tolosa Hunt syndrome (THS), also known as painful ophthalmoplegia, recurrent ophthalmoplegia, or ophthalmoplegia syndrome, is described as severe and unilateral periorbital headaches associated with painful and restricted eye movements. THS is one of the rare disorders recognized by the National Organization for Rare Disorders. The estimated annual incidence is one case per million per year.

Case Report: A 49-year-old male with a 12-year history of Type 2 Diabetes Mellitus presented with diplopia, ptosis, and right-sided hemifacial pain. Neurological examination revealed right-sided levator palpebrae superioris palsy, complete ophthalmoplegia, and conjunctival hyperemia. MRI confirmed orbital apex syndrome (OAS) in the right orbit known as Tolosa hunt syndrome (THS), while paranasal sinus HRCT showed chronic sinusitis with significant mucosal thickening and structural abnormalities. Management included high-dose intravenous methylprednisolone with gastroprotective therapy, followed by a tapering course of oral prednisolone and gabapentin for neuropathic pain.

Keywords: Tolosa Hunt Syndrome (THS), Orbital Apex Syndrome (OAS), Methylprednisolone, Ophthalmoplegia

INTRODUCTION

Tolosa Hunt syndrome, also known as painful ophthalmoplegia, recurrent ophthalmoplegia, or ophthalmoplegia syndrome, is described as severe and unilateral periorbital headaches associated with painful and restricted eye movements. THS is one of the rare disorders recognized by the National Organisation for Rare Disorders [2]. The estimated annual incidence is one case per million per year [4]. The syndrome of painful ophthalmoplegia consists of periorbital or hemi cranial 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]. THS was first described in the year 1954 by Dr. Eduardo Tolosa, a Spanish neurosurgeon [2]. Constant or episodic severe retro-orbital or periorbital pain is the initial feature of THS that usually resolves spontaneously but tends to relapse and remit. Double vision (diplopia) related to ophthalmoparesis or disordered eye movement occurs when cranial oculomotor (III), trochlear (IV), and abducens (VI) nerves are damaged by granulomatous inflammation [3]. THS is a

diagnosis of exclusion and it responds well to steroids [4].

The absence of a pathognomonic feature for THS necessitates its classification as a diagnosis of exclusion. A multitude of differential diagnoses associated with THS encompasses neoplasms, vasculitis, basilar meningitis, sarcoidosis, diabetes mellitus, and ophthalmoplegic migraine [5].

CASE STUDY

A 49-year-old male individual was admitted to the neurological department presenting with primary complaints of diplopia persisting for 15 days, accompanied by ptosis over the same duration, as well as right hemifacial pain. Furthermore, the patient has a documented history of Type 2 Diabetes Mellitus (T2DM) for the past 12 years, for which he is currently receiving insulin therapy, and there is an absence of any reported weakness in the limbs or episodes of fever.

Upon assessment utilizing the Glasgow Coma Scale (GCS), the subject exhibited a score of E4V5M6, which denotes a maximum response reflected by a cumulative score of 15, a finding that remained consistent at the time of discharge. During the systemic evaluation, the right pupil displayed sluggishness (2mm), while the left pupil demonstrated a reaction to light (2mm). The examination revealed the presence of right levator palpebrae superioris palsy, complete right eye ophthalmoplegia, and redness in the right eye, with Deep Tendon Reflex (DTR) assessments of the knees and ankles yielding negative results bilaterally.

When contrasting the MRI of the brain, which indicated the presence of right-sided orbital apex syndrome, there were no observable indicators of cavernous sinus thrombosis, no evidence of acute infarction, intraparenchymal hemorrhage, or intra-cranial space-occupying lesions, nor was there any abnormal lepto/pachymeningeal enhancement; moreover, the MRA of the circle of Willis and cervical vessels indicated the absence of significant vascular occlusion or stenosis. The HRCT of the paranasal sinuses revealed mucosal thickening in the bilateral maxillary sinuses, with the right side exhibiting greater involvement than the left, as well as sphenoid involvement being more pronounced on the left than the right, along with abnormal findings in the ethmoid and right frontal sinuses, which are suggestive of chronic sinusitis, accompanied by a deviation of the nasal septum to the left and bilateral inferior turbinate hypertrophy. The remainder of the neurological and systemic examination yielded unremarkable findings.

Blood investigation reports indicate that the hemoglobin level is measured at 10.9 g/dl, with a red blood cell count of 3.9 million cells per cubic millimeter, a hematocrit value of 32.7, and an erythrocyte sedimentation rate of 68 mm/hr. Biochemical analysis reveals that the total cholesterol level is 316.0 mg/dl, triglyceride concentration is 279 mg/dl, and low-density lipoprotein levels are recorded at 233.0 mg/dl.

The patient was treated with corticosteroids, specifically methylprednisolone at a dosage of 500 mg, administered as a glucocorticoid once daily in the morning in conjunction with the provision of gastroprotective therapy; upon discharge, the prednisolone 20 mg was given in addition with gabapentin 100mg night once daily for a week and follow up was suggested after a week to neurological department.

DISCUSSION

THS is a condition caused by nonspecific granulomatous inflammation of the cavernous sinus, superior orbital fissure and/or the apex of the orbit[6]. THS, also known as painful ophthalmoplegia, recurrent ophthalmoplegia, or ophthalmoplegia syndrome, is described as severe and unilateral periorbital headaches associated with painful and restricted eye movements[2]. 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 [7].

According to the International Headache Society Classification of 2004, THS is an entity that occurs rarely, its etiopathogenesis is unknown, it is manifested clinically by unilateral orbital pain associated with simple or multiple oculomotor paralyses, which resolves spontaneously but may recur. MRI orbital phlebography and biopsy are the recommended methods for making diagnosis [8].

The diagnostic criteria for the diagnosis of THS, defined by the 3rd edition of the International Classification of Headache Disorders (ICHD-3), include:[9]

A.	**Unilateral headache meeting criterion 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 third, fourth and / or fifth ipsilateral cranial nerves;
C.	**C. Evidence of causality demonstrated by the following two elements:**
1.	Headache preceded paresis of the third, fourth and / or sixth nerves of ≤ 2 weeks or developed with it;
2.	Headaches are localized around the eyebrow and the eye and
D.	**Not better explained by another diagnosis ICHD-3**

A 49-year-old male patient was admitted to the neurology department, reporting primary symptoms of diplopia that had persisted for 15 days, alongside ptosis over the same period, in addition to experiencing right hemifacial pain. During the comprehensive evaluation, the right pupil exhibited sluggish reactivity (2mm), whereas the left pupil demonstrated a light reflex (2mm). Clinical examination disclosed the presence of right levator palpebrae superioris palsy, complete ophthalmoplegia of the right eye, and conjunctival hyperemia in the right eye, with bilateral Deep Tendon Reflex (DTR) assessments of the knees and ankles returning negative results. Upon comparing the MRI of the brain, which indicated right-sided orbital apex syndrome, the HRCT of the paranasal sinuses revealed mucosal hypertrophy in the bilateral maxillary sinuses, with the right side showing more significant involvement than the left, and sphenoid involvement being more pronounced on the left side compared to the right. Additionally, abnormal findings in the ethmoid and right frontal sinuses were consistent with chronic sinusitis, accompanied by a leftward deviation of the nasal septum and bilateral hypertrophy of the inferior turbinates.

The patient underwent treatment with corticosteroids, specifically methylprednisolone, in conjunction with gastroprotective therapy.

CONCLUSION

Tolosa-Hunt Syndrome (THS) is an uncommon condition characterized by intense unilateral periorbital discomfort and limited ocular movements resulting from granulomatous inflammation affecting cranial nerves. It manifests with retro-orbital pain, diplopia, and ocular motor nerve impairments. As a diagnosis of exclusion, it necessitates differentiation from other conditions such as neoplasms, vasculitis, and ophthalmoplegic migraines. Although there is no definitive pathognomonic characteristic, the episodic nature of the syndrome and its favourable response to corticosteroid therapy are notable features. Timely diagnosis is essential to avert complications and to facilitate effective therapeutic intervention.

Declaration by Authors

Acknowledgement: None
Source of Funding: None
Conflict of Interest: The authors declare no conflict of interest.

REFERENCES

1. Kline LB. NOSOLOGICAL ENTITIES: The Tolosa-Hunt syndrome. Journal of Neurology, Neurosurgery & Psychiatry. 2001 Nov 1;71(5):577–82.
2. Chaitanya Amrutkar, Burton EV. Tolosa-Hunt Syndrome [Internet]. Nih.gov. StatPearls Publishing; 2019.
3. Zečević Penić S. Tolosa-Hunt Syndrome – Case Report. ACTA CLINICA CROATICA. 2017;331–7
4. K.C. S, K.C. S, Kunwar P, Dhungana K. Tolosa-Hunt Syndrome: A Case Report. Journal of Nepal Medical Association. 2021 Jul 1;59(238).
5. Maan Ali Almokdad, Ashraf ALakkad. Tolosa-Hunt Syndrome: A Case Report. Saudi Journal of Medical and Pharmaceutical Sciences [Internet]. 2023 Aug 9;9(08):534–8.
6. Paović J, Paović P, Bojković I, Nagulić M, Sredović V. Tolosa-Hunt syndrome--diagnostic problem of painful ophthalmoplegia. Vojnosanitetski pregled [Internet]. 2012 Jul;69(7):627–30.
7. Hunt WE, Meagher JN, LeFever HE, Zeman W. Painful ophthalmoplegia: Its relation to indolent inflammation of the cavernous sinus. Neurology [Internet]. 1961 Jan 1;11(1):56–6.
8. F İU, M Ö. Painful Ophthalmoplegia: A Case Report and Literature Review [Internet]. Agri: Agri (Algoloji) Dernegi'nin Yayin organidir = The journal of the Turkish Society of Algology. 2015.
9. Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia. 2013 Jun 14;33(9):629–808.

How to cite this article: Ajay R, Shubham B Harapanahalli, Veeranna Mohan Gadad, Gunupati Balamani Chandana, Gladys K Siji. Case report on Tolosa hunt syndrome in a diabetic patient: an uncommon presentation. *International Journal of Research and Review*. 2025; 12(1): 252-255. DOI: *https://doi.org/10.52403/ijrr.20250132*

******