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Case Report

Tolosa-Hunt Syndrome: A Rare Entity

Dr. Pillarsetty Pavan Kumar[1*], Dr. Uma M.A[2], Dr. Jagadish K.L[3], Dr. K P Jeswanth Kiran[4], Dr. Kotthinti Anil Kumar[5]

[1*]Post Graduate, Department of General Medicine, PES Institute of Medical Science & Research, Kuppam
[2]Professor and HOD of Medicine, Department of General Medicine, PES Institute of Medical Science & Research, Kuppam.
[3]Assistant Professor of Medicine, Department of General Medicine, PES Institute of Medical Science & Research, Kuppam.
[4]Post Graduate, Department of General Medicine, PES Institute of Medical Science & Research, Kuppam.
[5]Post Graduate, Department of General Medicine, PES Institute of Medical Science & Research, Kuppam.
Corresponding Author: Dr. Pillarsetty Pavan Kumar,
Email: dr.pillarsetty.pavan@gmail.com
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ABSTRACT
Tolosa-Hunt Syndrome (THS) is a rare disorder characterized by painful ophthalmoplegia due to idiopathic granulomatous inflammation in the cavernous sinus, superior orbital fissure, or orbital apex. The condition is often self-limiting but responds well to corticosteroid therapy. We present a case of a 54-year-old male with classic THS features, emphasizing the diagnostic challenges and management strategies. Early recognition and treatment are crucial to prevent morbidity associated with this condition.

Keywords: Tolosa-Hunt Syndrome, Painful Ophthalmoplegia, Cavernous Sinus, Cranial Neuropathy, Corticosteroids.

INTRODUCTION

Tolosa-Hunt Syndrome (THS) was first described in 1954 by Tolosa and later elaborated by Hunt et al. It is an uncommon condition with an estimated annual incidence of one case per million people.[1] THS is characterized by periorbital pain and ophthalmoplegia due to idiopathic granulomatous inflammation involving the cavernous sinus, superior orbital fissure, or orbital apex.[2] The clinical presentation may include exophthalmos, cranial nerve palsies (III, IV, VI), and sensory disturbances in the first (V1) and second (V2) divisions of the trigeminal nerve.[3] Recurrences occur in 40–50% of cases and can be ipsilateral, contralateral, or bilateral.[4] Diagnostic confirmation is challenging due to its overlap with other causes of painful ophthalmoplegia, requiring detailed neuroimaging and exclusion of other pathologies.[5] Corticosteroids remain the gold standard for treatment, with most patients demonstrating a dramatic response within 24–48 hours.[6]

Here, we report a case of THS in a 54-year-old male with characteristic clinical and radiological findings, highlighting the importance of early diagnosis and intervention.

Case Presentation

A 54-year-old male presented to the outpatient department with complaints of: Left periorbital pain for 16 days, described as throbbing and deep-seated, worsening over time. Double vision for 14 days, associated with blurred vision, redness, and mild swelling of the left eye.

History: The patient had recurrent headaches over the left frontal and temporal regions for two months, associated with photophobia, phonophobia, and nausea. He experienced similar episodes in the past month, relieved by topiramate and amitriptyline (Tryptomer). No known comorbidities prior to this episode. No history of smoking or alcohol consumption. Neurological Examination

Figure 1a: Left Lateral Rectus Palsy

Figure 1b: Left Medial Rectus Palsy

Higher mental functions: Normal. Visual acuity: Right eye: 6/12, Left eye: 6/24. Visual fields: Decreased. Extraocular movements: Left eye: Superior rectus (SR) and levator palpebrae superioris (LPS) spared; all other movements were lost. Right eye: Horizontal nystagmus present. Sensory loss: Over the V1 and V2 distributions of the trigeminal nerve. Pupils: Bilaterally normal with a reactive light reflex. Fundus examination: Grade II hypertensive retinopathy. Other systemic examinations: Unremarkable.

Laboratory Investigations

Complete blood count (CBC), renal function tests (RFT), liver function tests (LFT), thyroid profile: Normal. Erythrocyte sedimentation rate (ESR): 32 mm/hr (elevated). C-reactive protein (CRP): <5 mg/L. Urine sugar: 3+. HbA1c: 7.7% (Newly diagnosed Type 2 Diabetes Mellitus). Neuroimaging (MRI Brain & Orbit with Contrast) Enhanced lesion in the superior orbital fissure, with prominent cavernous sinus enhancement. Prominent superior ophthalmic vein, hazy intraconal fat, and minimal fluid along the optic nerve sheath (left side). Findings suggestive of Tolosa-Hunt Syndrome. Old infarcts in the left superior frontal and posterior parietal lobes, with tiny acute infarcts in the left posterior parietal lobes. Lacunar infarcts in the bilateral cerebral hemispheres.

Final Diagnosis

1. Tolosa-Hunt Syndrome
2. Newly Diagnosed Type 2 Diabetes Mellitus

Management

Intravenous Methylprednisolone (500 mg/day) for 2 days followed by Oral Prednisolone (40 mg/day) with gradual tapering Ecosprin-AV (75/20 mg) once daily Oral hypoglycemic agents (OHAs) initiated for diabetes management

The patient showed marked clinical improvement within 48 hours of steroid therapy, confirming the diagnosis of THS.

DISCUSSION

THS can affect individuals of any age, typically between the 2nd and 8th decades of life, without significant gender predilection.[7] the syndrome is characterized by recurrent periorbital pain, headaches, and ocular motor cranial nerve palsies.[8] the third, sixth, fifth, and fourth cranial nerves are involved in descending order of frequency.[9]

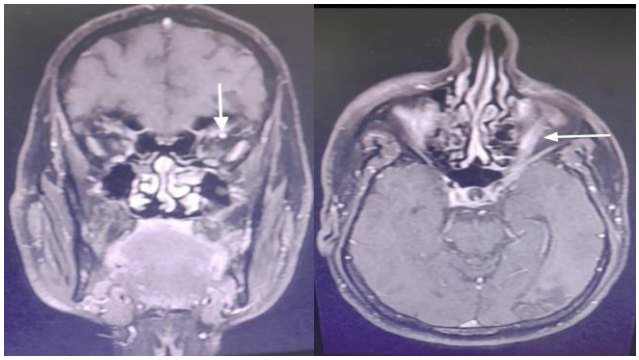

Figure 2a: Prominent Superior Ophthalmic Vein on Left Side

Figure 2b: Hazy Intracranial Fat and Minimal Fluid along the Optic Nerve Sheath on Right Side

Diagnostic Criteria

According to the International Headache Society (IHS) 2004, the diagnosis of THS requires:

1. Periorbital pain and ophthalmoplegia affecting cranial nerves III, IV, or VI.
2. Granulomatous inflammation on MRI or biopsy.

3. Exclusion of other causes of painful ophthalmoplegia.[10]

The sensitivity of these criteria is 95–100%, but specificity remains low (50%) due to overlap with conditions such as cavernous sinus thrombosis, meningioma, pituitary apoplexy, and carotid-cavernous fistula.[11]

Classification of THS: Based on neuroimaging findings, THS can be categorized as: Benign: No abnormal MRI findings, Inflammatory: MRI or biopsy shows granulomatous inflammation and Symptomatic: A specific lesion is detected, such as neoplasms or vascular abnormalities.[12]

Management & Prognosis

High-dose corticosteroids remain the mainstay of treatment, leading to rapid symptom resolution.

Most cases respond within 24–48 hours, although relapses occur in 40–50% of patients.[13]

Long-term steroid therapy is sometimes required in refractory or recurrent cases.

Alternative immunosuppressants like methotrexate or azathioprine may be considered in steroid-resistant cases.[14]

CONCLUSION

Tolosa-Hunt Syndrome is a rare but treatable cause of painful ophthalmoplegia, requiring early diagnosis and intervention to prevent complications. While unilateral cases are common, bilateral or alternating involvement is extremely rare and should prompt careful neuroimaging assessment. Immediate steroid therapy remains crucial, as seen in our patient who demonstrated a dramatic response.

REFERENCES

1. Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38(1):1-211.
2. La Mantia L, Curone M, Rapoport AM, Bussone G. Tolosa–Hunt syndrome: critical literature review based on IHS 2004 criteria. Cephalalgia. 2006;26(7):772-81.
3. Hung CH, Chang KH, Wu YM, Chen YL, Lyu RK, Chang HS, et al. A comparison of benign and inflammatory manifestations of Tolosa-Hunt syndrome. Cephalalgia. 2013;33(11):842-52.
4. Hao R, He Y, Zhang H, Zhang W, Li X, Ke Y. The evaluation of ICHD-3 beta diagnostic criteria for Tolosa-Hunt syndrome: A study of 22 cases of Tolosa-Hunt syndrome. Neurol Sci. 2015;36(5):899-905.
5. Jain R, Sawhney A, Koul RL, Chand P. Tolosa-Hunt syndrome: series of six cases. Indian J Ophthalmol. 2003;51(4):335-8.
6. Kline LB, Hoyt WF. The Tolosa-Hunt syndrome. J Neurol Neurosurg Psychiatry. 2001;71(5):577-82.
7. Smith JL, Taxdal DS. Painful ophthalmoplegia. The Tolosa-Hunt syndrome. Am J Ophthalmol. 1966;61(6):1466-72.
8. Cohen-Inbar O, Lee CC, Xu Z, Da Silva I, Schlesinger D, Sheehan JP. Gamma knife radiosurgery for idiopathic granulomatous chronic orbital apex syndrome: a case report and review of the literature. Surg Neurol Int. 2014; 5:171.
9. Keane JR. Painful ophthalmoplegia: the Tolosa-Hunt syndrome. Clin Neurosurg. 1980; 27:593-602.
10. Younge BR, Gorman CA, Garrity JA, Cross SA. Ophthalmoplegia and facial pain. The Tolosa-Hunt syndrome. Ophthalmology. 1990; 97(5):680-9.
11. Saeed P, Rootman J, Nugent RA, White VA, Heran MK, Graeb DA. Painful ophthalmoplegia: clinical and imaging diagnosis of Tolosa-Hunt syndrome. Ophthalmology. 2003;110(4):718-23.
12. Schuknecht B, Sturm V. Imaging in Tolosa-Hunt syndrome. Neuroradiology. 2009;51(9):633-40.
13. Miller NR. Diagnosis and management of Tolosa-Hunt syndrome. J Neuroophthalmol. 2002;22(1):68-9.
14. Matsuo T, Fukui M, Okeda R. Tolosa-Hunt syndrome: a clinicopathological study. J Neurol Neurosurg Psychiatry. 1988;51(11):1512-5.