Case Report

A rare presentation of Tolosa – Hunt syndrome in a young male

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ABSTRACT

The primary care provider, nurse practitioner, and internist often encounter patients with a headache. However, when they encounter a patient with an intense headache and ophthalmoplegia, the care plan no longer resides with a general practitioner, and referral to a neurologist becomes mandatory. This is due to the vast differentials involved and rare pathologies that one may come across during their medical practice. One such case of a 45-year-old male with similar complaints encountered during a routine emergency day led to a diagnosis of a rare neuro-immunological disorder, possibly a diagnosis of exclusion.

Key words: Cavernous sinus, Glucocorticoids, Magnetic resonance imaging, Ophthalmoplegia, Tolosa-Hunt syndrome

Tolosa–Hunt syndrome (THS) is a rare cause of painful ophthalmoplegia characterized by non-specific granulomatous inflammation involving the cavernous sinus, superior orbital fissure, or orbital apex, and typically presents with unilateral orbital or periorbital pain associated with paresis of the third, fourth, and/or sixth cranial nerves. It is recognized as a rare disorder by major rare disease registries and classified among painful cranial neuropathies in contemporary headache classifications, underscoring its low incidence and the need for a high index of suspicion in clinical practice. THS remains a diagnosis of exclusion in which clinical presentation, supportive neuroimaging, and a characteristic, often dramatic, response to corticosteroid therapy together guide diagnosis in the absence of a specific biomarker [1-4].

Despite its stereotypical description, reported cases demonstrate considerable heterogeneity in age at onset, pattern of cranial nerve involvement, imaging appearances, and clinical course, including the risk of relapse and the occasional need for steroid-sparing immunosuppression. Such variability can make early recognition challenging and may delay appropriate treatment, particularly when presentations are incomplete, atypical, or overlap with more common vascular, infectious, or neoplastic causes of cavernous sinus syndrome. Case reports and series, therefore, remain an important source of information to refine diagnostic criteria, highlight imaging clues, and illustrate therapeutic responses in real-world settings [5,6].

We report a middle-aged male with painful ophthalmoplegia and isolated involvement of the ophthalmic division of the trigeminal nerve with fourth and sixth cranial nerve palsies, in whom extensive evaluation excluded alternative etiologies and magnetic resonance imaging (MRI) demonstrated inflammatory changes in the cavernous sinus, with rapid and sustained improvement following high-dose corticosteroid therapy. This case is being reported to add to the limited literature on THS in relatively young males, to emphasize the diagnostic value of correlating segmental cranial nerve involvement with targeted cavernous sinus imaging, and to reinforce that early steroid therapy can be both diagnostic and therapeutic, potentially preventing permanent cranial nerve dysfunction.

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CASE REPORT

A 45-year-old male, laborer by occupation, presented with a complaint of right-sided frontal headache for 6 months, which was insidious in onset, non-progressive, throbbing in nature, and present throughout the day. The pain was not associated with photophobia, phonophobia, or any routine activity, and there was no diurnal variation. He also complained of right-sided periorbital tingling sensation for the last 5 days, drooping of the right eyelid, and vertical diplopia for the last 2 days. He gave no history of trauma or similar complaints in the past. He was a non-hypertensive and a non-diabetic. However, he is an occasional alcoholic and a chronic bidi smoker.

The patient was conscious, co-operative, and well-oriented to time, place, and person. His pulse rate was 86 beats/min, good volume, and equal on both sides. His blood pressure was 110/70 mm Hg, measured on the right brachial artery, in sitting position. There were no

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signs of pallor, cyanosis, clubbing, edema, or palpable lymphadenopathy. On ocular examination (Figs. 1 and 2), the patient's right eye was partially closed during the first visit, with an upper lid margin to skin crease distance of 2 mm on the right and 6 mm on the left. His visual acuity (Snellen Chart) was 6/12 in the right eye and 6/6 in the left eye, with no diminution in color vision. There were no signs of proptosis, chemosis, or ocular discharge. Pupils were normal in size, and pupillary light (direct and consensual) and corneal reflexes were present bilaterally. However, the patient had a tingling sensation in the right periorbital region (Ophthalmic division – Vth Cranial Nerve). On extraocular motility assessment, the patient was unable to abduct and intort his right eye (IVth and VIth Cranial Nerve Palsies). Examination of the left eye was normal with no fundus changes bilaterally. Examination of other cranial nerves was normal. Other systemic examinations were normal.

Laboratory investigations of the patient are shown in Table 1. MRI of the brain and orbits with contrast revealed an ill-defined, extra-axial soft-tissue lesion along the lateral wall of the right cavernous sinus extending anteriorly toward the orbital apex, partially encasing the cavernous segment of the internal carotid artery (ICA). The lesion appeared iso- to hypointense relative to gray matter on T1-weighted images and showed variable signal intensity on T2-weighted sequences, with areas of intermediate to mildly hyperintense signal suggestive of inflammatory tissue rather than a highly cellular neoplasm. Following gadolinium administration, there was homogeneous and intense contrast enhancement, and subtle narrowing and irregularity of the cavernous ICA lumen were noted, consistent with periarteritic involvement. These imaging features, in the appropriate clinical context, favored an inflammatory etiology compatible with THS and helped exclude alternative causes such as neoplastic, infectious, or thrombotic cavernous sinus pathology.

The temporal relationship between unilateral orbital pain and the subsequent onset of ipsilateral cranial nerve palsies, together with later imaging-confirmed involvement of the cavernous sinus, fulfilled the International Classification of Headache Disorders, 3rd edition diagnostic criteria for THS as defined by the International Headache Society.

Clinical symptoms and signs initially raised concern for several causes of painful ophthalmoplegia, including intracavernous aneurysm, diabetic microvascular neuropathy, periarteritis nodosa, parasellar neoplasm or infection, cavernous sinus thrombosis, carotid–cavernous fistula, and THS. In view of India's high infectious burden, tuberculous pachymeningitis and tuberculoma, invasive fungal infections, and inflammatory conditions such as sarcoidosis were also considered, along with septic cavernous sinus thrombosis. However, the absence of systemic features of infection, normal inflammatory markers, negative serological and CSF studies, and MRI findings showing a homogeneously enhancing cavernous sinus lesion without evidence of abscess, meningeal thickening, venous thrombosis, or destructive skull base changes made these alternative diagnoses unlikely, leaving THS as the most compatible explanation for the patient's presentation.

With the suspicion of THS at the back of our minds, the patient was given Injectable Methyl Prednisolone 500 mg for 3 days (Pulse therapy), following which

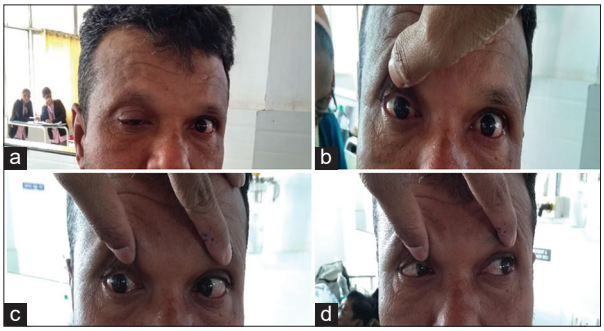

Figure 1: (a) Right eye ptosis; (b) on looking straight ahead; (c) on right lateral rotation; (d) on left lateral rotation

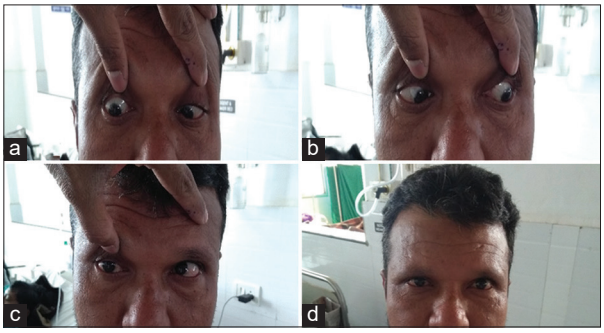

Figure 2: Right eye ptosis (a) on depression and right lateral rotation; (b) on depression and left lateral rotation; (c) on elevation and right lateral rotation; (d) 72 h since initiation of steroid therapy

Table 1: The laboratory parameters of the patient

Laboratory parameter	Normal range	Observed value
Hemoglobin	12–16.2 g/dL	14.5
Total leucocyte count	3540–9060/cmm	7620
Platelet count	165000–415000/cmm	195000
Hematocrit	35.4–46.4	40.1
Mean corpuscular volume	80–96 fl	85.3
Urea	7–20 mg/dL	26
Creatinine	0.5–1.2 m/dL	0.8
Serum Sodium	136–146 mEq/L	140
Serum Potassium	3.5–5.5 mEq/L	4.8
Erythrocyte sedimentation rate	0–15 mm/hr	4
C-reactive protein	<3 mg/L	2.5
Lipid profile		
Serum cholesterol	150–200 mg/dL	172
HDL cholesterol	40 –60 mg/dL	44
LDL cholesterol	70–140 mg/dL	122
Serum triglycerides	10–150 mg/dL	96
HIV, HBsAg, HCV	Non-reactive	
Antineutrophilic antibody	Negative	
CSF IgG	Negative	

the patient showed signs of improvement with gradual resolution of right eye ptosis.

After 72 h of initiating steroid therapy, the headache subsided (Fig. 2d). The patient was eventually started on Oral Prednisolone (1 mg/kg), thereafter tapered weekly. Patient followed up in the Medicine outpatient department after 15 days, and his visual acuity in the right eye improved to 4/6.

DISCUSSION

Painful ophthalmoplegia secondary to idiopathic granulomatous inflammation of the cavernous sinus or the superior orbital fissure that is steroid responsive was given the eponym THS by Smith and Laxdal [3] in 1966 when they reported five cases of painful ophthalmoplegia and confirmed the importance of therapeutic trials of steroids as a diagnostic test.

Proposed mechanisms for THS include an idiopathic, non-specific granulomatous inflammatory process involving the walls of the cavernous sinus and adjacent structures, leading to painful dysfunction of the oculomotor, trochlear, and abducens nerves. A possible autoimmune basis has been suggested, with localized immune-mediated inflammation targeting periarterial tissues and dural structures, which would explain the typical steroid responsiveness and tendency to relapse in a subset of patients. Direct involvement of cavernous sinus structures, including cranial nerves and the ICA, likely accounts for the characteristic combination of orbital pain, ophthalmoplegia, and occasional vascular imaging abnormalities seen in THS [6,7].

THS is an uncommon but important cause of painful ophthalmoplegia, and much of the available evidence comes from single-center series and case reports, including several from India. Indian reports describe patients across a wide age range, often presenting with unilateral orbital pain, cranial nerve III, IV, or VI palsies, and characteristic cavernous sinus or orbital apex enhancement on MRI, similar to the present case [5,6]. These series highlight that THS is likely under-recognized, particularly in resource-limited and rural settings where access to dedicated neuroimaging and subspecialty care may be restricted, leading to under-reporting in the literature [7].

Glucocorticoids remain the cornerstone of treatment, with rapid pain relief and gradual improvement in cranial nerve deficits reported in most series. Typical regimens include high-dose intravenous methylprednisolone pulses followed by oral prednisolone, or high-dose oral steroids alone, tapered slowly over several weeks to months to minimize relapse [8-10]. In our patient, intravenous methylprednisolone pulse therapy followed by a tapering course of oral steroids produced brisk resolution of pain within 72 h and progressive improvement in ophthalmoplegia, in keeping with prior reports. For patients with recurrent disease or those in whom prolonged steroid exposure is undesirable, steroid-sparing immunosuppressive agents such as azathioprine, mycophenolate mofetil, methotrexate, or cyclophosphamide have been used as adjuncts or maintenance therapy [9,10].

Long-term outcome studies indicate that, although the initial response to corticosteroids is usually excellent, recurrence is not uncommon. Recurrence rates in published series range roughly from 30% to 50%, with relapses occurring months to years after the index episode, sometimes on the same side and occasionally contralaterally. Despite this, the overall prognosis is generally favorable, with most patients achieving good functional recovery and only a minority left with persistent cranial nerve deficits or residual ophthalmoplegia. The largest single-center Indian series demonstrated that the use of steroid-sparing agents was associated with a lower recurrence rate compared with steroids alone, suggesting a potential role for early introduction of immunosuppressants in patients at high risk of relapse [9-12].

From an Indian perspective, accurate diagnosis of THS is particularly challenging because of the need to rigorously exclude more common infectious and inflammatory mimics such as tuberculous pachymeningitis, invasive fungal disease, sarcoidosis, and septic cavernous sinus thrombosis, all of which are highly relevant in this epidemiological setting [13]. Our patient underwent detailed systemic evaluation and neuroimaging that failed to reveal evidence of infection, granulomatous systemic disease, or venous thrombosis, and instead showed imaging changes confined to the cavernous sinus with a typical steroid-responsive course, supporting the diagnosis of THS. Nevertheless, current data largely derive from single-center experiences, and there is a clear need for multicentric, prospective studies in India and other low- and middle-income countries to better define the true burden, predictors of recurrence, and optimal use of steroid-sparing therapy [12,13]. Such collaborative efforts would also help to address likely under-reporting from rural and peripheral centers, thereby improving recognition and long-term outcomes in patients with this rare but treatable cause of painful ophthalmoplegia.

CONCLUSION

THS should be considered in patients presenting with unilateral painful ophthalmoplegia once more common causes have been excluded, but confirmation increasingly depends on timely, targeted neuroimaging of the cavernous sinus and orbital apex. Early MRI not only supports the diagnosis but also helps rule out infectious, vascular, and neoplastic mimics that are particularly relevant in high-burden settings. Greater awareness of this entity among primary care physicians and emergency practitioners is essential so that appropriate referrals, imaging, and steroid therapy are not delayed, thereby reducing the risk of permanent cranial nerve deficits.

Establishing a national registry or coordinated reporting system for THS, especially incorporating data from rural and peripheral centers, would provide much-needed epidemiological and outcome data, guide treatment strategies, and improve recognition of this rare but highly treatable cause of painful ophthalmoplegia.

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