Case Study

Clinical Images and Medical Case Reports

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A Case of A Rare Unilateral Headache: A Clinical Case Study

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Received: December 16, 2024; **Accepted:** January 06, 2025; **Published:** January 12, 2025

Abstract

Purpose: To report a rare case of unlilateral headache and ophthalmopelgia and its management. The aim of our report is to raise awareness among clinicians about Tolosa-Hunt syndrome as a potential cause of unilateral headaches with symptoms of cranial nerve involvement.

Methods: A 42-year-old man with a history of severe, constraining headaches had subsequent eye movement difficulties. His symptoms quickly escalated to include double vision and restricted eye movement, compounded by significant swelling and erythema around his eye. A brain and orbital MRI (Magnetic Resonance Imaging) revealed significant thickening of the left cavernous sinus, in addition to pachimeningeal thickening in the left Meckel's cave and nearby dural thickening.

Results: The patient exhibited dysfunction of the left optic (II), abducens (IV), oculomotor (III), trochlear (IV), ophthalmic branch of the trigeminal nerve (V), and MRI abnormalities that satisfy the diagnostic criteria for THS. The patient was prescribed prednisolone, which significantly improved headache and ptosis after 72 hours, and cranial nerve palsies improved within two weeks. After a year, no headaches or ophthalmia recurred.

Conclusion: This case report describes an infrequent presentation and is important because the symptoms of this syndrome can be similar to those of other clinical conditions, and a definitive diagnosis can only be made through appropriate imaging or a biopsy. The need to get biopsy or MRI data before confirming a diagnosis of THS has been emphasized by the most recent guidelines.

Keywords: Headache, Painful ophtalmoplegia, Tolosa-hunt syndrome, Case reports

Introduction

Tolosa-Hunt syndrome (THS) is characterized by an intense and unilateral periorbital headache, accompanied by painful and limited eye movements (1).

The idiopathic inflammation results in external compression of the neurovascular structures that pass through the cavernous sinus or orbital apex (2). Diagnostic criteria include headache, cavernous sinus inflammation, superior orbital fissure, confirmed by MRI or biopsy, and paralysis of ipsilateral cranial nerves III, IV, or VI (3).

The incidence of THS is estimated to be approximately one case per million per year (4).

According to the National Organization for Rare Disorders, the mean age at which symptoms first appear is 41 years. While typically occurring on one side, it is possible for both sides to be impacted (about 5%). No predisposition exists between males and females (5).

The diagnosis of THS is mostly based on the process of exclusion, where other conditions such as vascular, neoplastic, infective, and inflammatory diseases that cause

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Citation: Bhadorizadeh L (2025) A Case of A Rare Unilateral Headache: A Clinical Case Study. Clin Img and Med Case Rep Vol.2 No.1.

painful ophthalmoplegia are ruled out with appropriate examinations.

Herein we report a case of a 42-year-old man who presented with headache, ophtalmoplegia, cunjunctive injection, and eyelid swelling. It's worthy to note that this was a challenging case due to several indefinite differential diagnoses, including THS, Cerebral Sinovenous Thrombosis (CSVT), and orbital cellulitis, that require different treatment.

Case Report

We report the case of a 42-year-old male who presented to our emergency department with a sudden-onset left-sided headache and an ipsilateral painful eye movement. He had a history of left frontal and retroorbital headaches 7 days prior to admission (Figure 1).

The headache exhibited a severe, continuous, constrictive, non-throbbing nature. It frequently disrupted the patient's sleep and, as it progressed, didn't show a favorable response to analgesic medications. Four days after the headache, the patient experienced diplopia, intermittent blurred vision, and restricted eye movement, accompanied by left eyelid drooping, swelling, erythema around the eye, and conjunctivitis. The patient didn't report experiencing any symptoms of nausea, vomiting, visual auras, photophobia, or phonophobia. The patient was on asthma medication, and other clinical history was unremarkable.

At admission, the patient was afebrile, had rhinorrhea and nasal congestion, and vital signs were within normal range. The ophtalmologic examination revealed left upper lid ptosis and restricted left eye movement in all directions, indicating the involvement of cranial nerves III, IV, and VI. Additionally, a reduced sensation was observed in the region innervated by the left ophthalmic branch of the trigeminal nerve. The pupils exhibited normal size and reacted appropriately to light stimulation. His visual acuity in the right and left eyes was 20/20 and 20/30, respectively, and funduscopic examination was normal (Figures 2A-D). Laboratory findings were unremarkable except for elevated C-reactive protein levels (23 mg/dL, normal range in adults: 0.3–1). Further serological and autoimmune

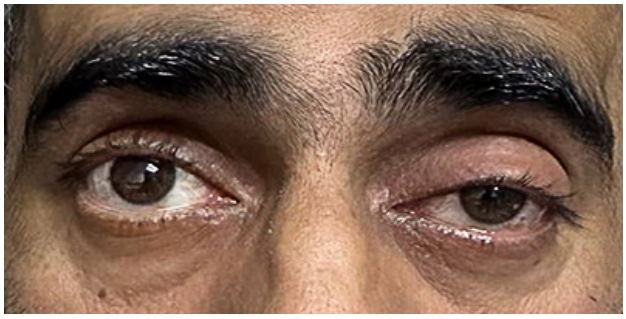

Figure 1:

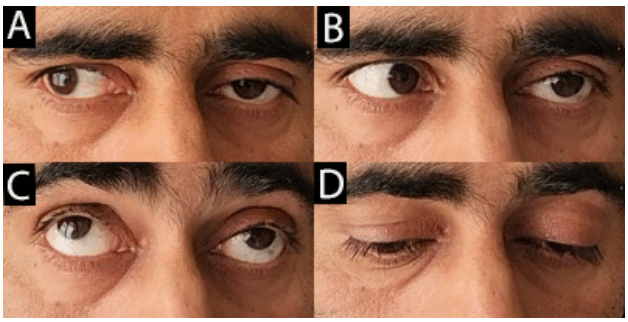

Figure 2:

panels were within the normal range except for increased serum IgG (3782 mg/dL; normal range in adults: 600–1700 mg/dl) levels. Also, the cerebral spinal fluid was found to be normal. The patient's primary presentation was best explained by CSVT, or orbital cellulitis; therefore, anticoagulation and emperical antibiotic therapy were started. However, in the course of treatment, the Magnetic Resonance Venography investigation didn't confirm the CSVT diagnosis, and no improvement in clinical manifestations was attained. Contrast-enhanced MRI of the brain and orbits demonstrated a remarkable enhancement in the left cavernous sinus with nearby dural thickening and also pachimeningeal thickening in the left Meckel's cave. There is evidence of thickening of the pachymeninges around the optic nerve, but there are no signs of optic neuritis (Figures 3A-B).

By exclusion of other critical conditions, prednisolone was initiated at a dose of 1 mg/kg body weight. After 72 hours of steroid therapy, the patient's headache and ptosis significantly improved. The findings were consistent with the THS diagnosis. Prednisolone was continued for 2 weeks and then tapered to discontinue. Cranial nerve palsies improved gradually within two weeks. After 1 year of follow-up, the patient didn't report any recurrence of headaches or ophthalmia.

Discussion

According to the International Classification of Headache Disorders 3rd edition-beta, THS is characterized by unilateral orbital pain associated with paresis of one or more of the third, fourth, and/or sixth cranial nerves caused by granulomatous inflammation in the cavernous sinus, superior orbital fissure, or orbit. Headache either develops concurrently with or two weeks prior to cranial nerve paresis. Some documented cases of TSH have demonstrated additional engagement of the trigeminal nerve (often the first division) or the optic nerve (6).

The present patient had impairment of the left optic (II), abducens (IV), oculomotor (III), trochlear (IV), ophthalmic branch of the trigeminal nerve (V), and MRI abnormalities that meet the diagnostic criteria for THS.

Hao et al. studied 22 THS patients and found that cranial nerve palsy was most common in the third (91%),

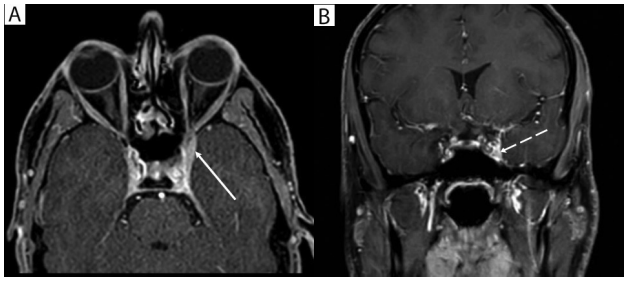

Figure 3:

fourth (91%), sixth (68%), ophthalmic and maxillary branches of the fifth cranial nerve (18% and 9%), and optic nerve (9%) (7).

MRI alone for diagnosis is problematic as it may reveal any aberrant tissue, potentially leading to false positives due to neoplasms, inflammatory diseases, and infections. THS is a diagnosis of exclusion and requires a comprehensive investigation (3).

The latest guideline suggestion about the role of MRI in diagnosis has been stricter, changing from the requirement of 'and/or demonstration of granulomas by MRI or biopsy' in ICHD-II to the requirement of 'demonstrated by MRI or biopsy' in ICHD-III beta (6-8). According to ICHD-III beta, patients with normal MRI are no longer labeled as THS unless a biopsy shows evidence of granolumatous inflammation.

Zhang et al. found that only 24 out of 46 patients with THS (52.2%) exhibited MRI abnormalities (9). There have been concerns about the ICHD-III beta diagnostic criteria's sensitivity and specificity (3-9).

The invasive procedure of obtaining pathological samples from THS lesions, often found near the skull base, poses numerous risks. Thus, MRI has gained importance.

The ICHD-III beta revised the significance of glucocorticoids in the identification of THS; however, it continues to include them in the diagnostic criteria (10).

After 72 hours of administering glucocorticoids, our patient showed complete pain relief and partial cranial nerve palsy improvement. It was consistent with the Zang et al. study, which reported 40% complete and 60% partial pain relief within 72 hours and 70% partial nerve palsy relief within 72 hours.

THS is a rare disease that causes painful ophthalmoplegia due to idiopathic granulomatous inflammation of the cavernous sinus. Our objective is to add to the existing body of data concerning the course and results of this disease process, which is currently lacking.

Conflict of Interest

The authors declare no conflict of interest.

Funding

No funding was received for this study.

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