Case Report

Tolosa-Hunt syndrome; a diagnosis of exclusion: A case report

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Abstract

Tolosa-Hunt syndrome is a rare cause of unilateral headache with painful ophthalmoplegia. Only a limited number of cases are reported in the literature. We present a case of Tolosa-Hunt syndrome in a middle-aged male who presented to the Teaching Hospital, Anuradhapura.

A diagnosed patient with hypertension presented with gradual onset right-sided headache and diplopia for five days duration. On examination, there were right-sided palsies of cranial nerves III, IV, V-1, and VI . Magnetic Resonance Imaging (MRI) of the brain with contrast was unremarkable. The patient was treated with steroids after excluding other possible causations of painful ophthalmoplegia. A dramatic improvement to steroids was observed.

Tolosa-Hunt syndrome responds well to steroid therapy even though there is no proven benefit of steroids to reduce recurrence rates. Careful evaluation needs to be carried out to rule out other possible causes of similar clinical and radiological features as they may be masked by steroid therapy.

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Introduction

The characteristic clinical presentation of the Tolosa-Hunt syndrome (THS) was first described in the year 1954 by Dr. Eduardo Tolosa, a Spanish neurosurgeon [1]. After similar cases being reported by Hunt et al. in 1961 [2], Smith and Taxdal named it THS in 1966 [3]. It typically presents with orbital pain associated with the palsy of the third, fourth, or sixth cranial nerves [4]. The condition occurs due to inflammation of the cavernous sinus and surrounding structures and is usually idiopathic [5]. In some uncommon cases, dysfunction of the optic, trigeminal, facial, acoustic nerve, and sympathetic innervation of the pupil can be observed [6,7] as in the discussed patient where he had cranial nerve V-1 involvement.

Case report

A 48-year-old male was admitted with right-sided, gradual onset, persistent, and worsening retro-orbital pain for five days. He also complained of diplopia for three days. The headache was not associated with vomiting or fever. There was no tearing, reddening of the eyes. He remained conscious and alert. There was no history of trauma to the head in the recent past. He had no similar episodes in the past. His past medical history

was unremarkable. The patient was a non-smoker and a teetotaler.

On physical examination, the patient was hypertensive (blood pressure 150/84 mmHg), and the other vital signs were within the normal range. His Glasglow Coma Scale (GCS) score was 15 out of 15. There were no signs of meningism. His pupils were equal with intact pupillary reflexes. He had a squint with right-sided palsies in cranial nerves IV, V-1, and VI. Fundi were bilaterally normal. There were no other neurological deficits. Two days later, he developed partial ptosis of the right eye. However, the right pupil remained normal (Figure 1).

Figure 1: Examination of eyes (pre-treatment); palsies of right-sided cranial nerves III, IV and VI

Urgent non-contrast computed tomography (CT) scan of the head was unremarkable. His full blood count, C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), fasting blood sugar, serum glycosylated haemoglobin (HbA1c), serum sodium, potassium and calcium levels, liver profile, urine full report, and renal functions were within the normal range. The contrast magnetic resonance imaging (MRI) scan of the brain with cavernous sinus and orbital cuts was unremarkable. Fluorescent treponemal antibody test, antinuclear antibody, serum protein electrophoresis, and antinuclear cytoplasmic antibody were negative. Angiotensin-converting enzyme level could not be performed as the testing facility was unavailable in the hospital. A presumptive diagnosis of THS was made.

He was started on oral prednisolone 60 mg daily. Within 24 hours, his headache was completely resolved. Over the next three days, there was a marked improvement of the cranial nerve palsies and ptosis, and the diplopia gradually improved. On review after two weeks of steroid therapy, he was fully recovered with normal eye movements (Figure 2). His steroid therapy was tailed off for over one month. He remained well at the six-month review.

Discussion

This report presents a case of THS, arare disease with an estimated annual incidence of one case per million per year [8]. It is caused by a nonspecific inflammatory process in the cavernous sinus with possible extension into the supraorbital fissure and orbital apex. No information is available as to what triggers the inflammatory process [8]. Except for its remitting-relapsing nature, it is a relatively benign disorder.

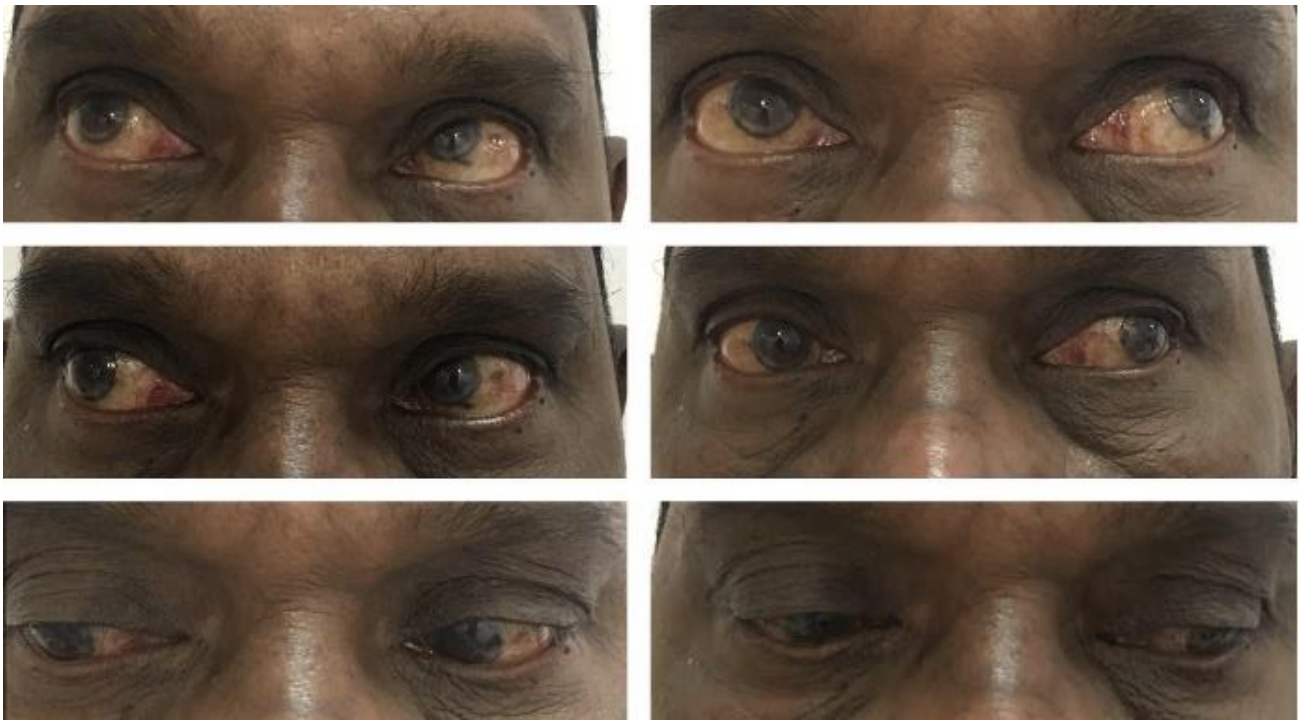

Figure 2: Examination of eyes (post-treatment); restored functions of right-sided extraocular muscles

A wide range of pathologies may produce clinical features similar to THS. These include craniocerebral trauma, various vascular causes, neoplastic processes, and inflammatory causes due to infective or non-infective processes [8]. Thus, THS is a diagnosis of exclusion. Gadolinium-enhanced MRI is the imaging modality of choice to evaluate THS, where it can show thickening of the cavernous sinus because of the presence of abnormal soft tissue swelling, which is isointense on T1, iso or hypointense on T2 and enhances with contrast. Other MRI findings include abnormal enlargement and enhancement of the cavernous sinus extending through the superior orbital fissure into the orbital apex. The major limitation of MRI findings in THS is their lack of specificity [9]. Yet, MRI helps to exclude other common lesions involving the cavernous sinus [10]. The patient described did not have any identifiable secondary cause from the history, examination, and investigations. Corticosteroids remain the mainstay of treatment for THS [2]. Spontaneous remission is also known to occur. Rapid amelioration of pain within 24–48 hours with the administration of steroids is typical and supports the diagnosis, as was observed in our patient. However, many of the mimics, including chordoma, giant cell tumour, lymphoma, and epidermoid, also respond rapidly to corticosteroids [8], highlighting that imaging is important to exclude these cases before starting on steroids.

Conclusion

THS is a rare yet treatable cause of painful ophthalmoplegia. Given close similarities of THS to its mimics and the inability to perform a biopsy as its risks outweigh the benefits, diagnosis of THS is by careful clinical and radiological exclusion of its mimics. Attending clinicians need to re-examine the diagnosis during the follow-up to avoid missing sinister pathologies and recurrence.

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