Case Report

Painful ophthalmoplegia due to Tolosa-Hunt syndrome: a case report

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Abstract

Tolosa-Hunt syndrome (THS) is a rare clinical entity, characterized by sudden onset of painful ophthalmoplegia and prompt response to steroid therapy. Generally, it involves the third, fourth, and sixth cranial nerves due to presence of non-specific inflammation at the level of cavernous sinus or superior orbital fissure. In some cases, first or second division of trigeminal nerve may also be involved. Here we present a case report on THS, a rare cause of painful ophthalmoplegia. The patient presented with sudden onset, unilateral headache, drooping of left upper eyelid followed by diplopia. On examination, the patient had complete ophthalmoplegia, along with ophthalalmic division of trigeminal nerve was also involved. Magnetic resonance imaging (MRI) brain showed expansion of left cavernous sinus suggesting THS. Other differentials of THS were ruled out on the basis of careful history, examination, and investigations.

KEY WORDS: Painful ophthalmoplegia, ptosis, diplopia, Tolosa-Hunt syndrome, cavernous

Introduction

The Tolosa-Hunt syndrome (THS) is characterized by painful ophthalmoplegia that is steroid responsive. THS is a rare neurologic disorder with an estimated incidence of one case per one million of populations.[1]

In 1954, Tolosa[2] reported the first patient with this syndrome, who presented with left orbital pain, ipsilateral progressive visual loss, total left ophthalmoplegia, and reduced sensation over the first division of the trigeminal nerve. After 7 years of this, in 1961, Hunt et al.[3] described this clinical entity, on the basis of six patients. It is an idiopathic condition which develops due to non-specific granulomatous inflammatory process in the region of the cavernous sinus/superior orbital fissure.

Although it is considered a benign condition, permanent neurologic deficits can occur, and relapses can happen often requiring prolonged immunosuppressive therapy. THS is a diagnosis of exclusion and it must be carefully differentiated from more malignant causes of cavernous sinus involvement and painful ophthalmoplegia.

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Case Report

A 28-year-old woman presented to the medicine casualty with sudden onset of left sided headache and drooping of the left upper eyelid. After about 1 week of this, she developed double vision. Headache was constant, unilateral left sided, periorbital extending retroorbitally into the frontal and temporal regions, and of severe intensity. There was no history of vision loss. Past history was not significant. The patient was afebrile on presentation, with a pulse rate of 86 beats/min and a blood pressure of 124/80 mmHg. Rest of the general physical examination was normal.

There were no signs of pallor, icterus, cyanosis, clubbing, raised jugular venous pressure, and pedal edema. She was conscious and oriented and other higher mental functions were normal. On ocular examination, left sided ptosis was present (Figure 1). Patient had dilated non-reactive left pupil on presentation. Both direct and indirect pupillary reflexes were absent on left side initially but with start of steroid therapy, pupil became mid dilated and started sluggishly reacting to light.

Patient had complete ophthalmoplegia on left side at presentation. But with start of steroid therapy, fourth cranial nerve palsy improved partially whereas third and sixth cranial nerve palsy persisted on left side (Table 1, Figure 2).

Right eye examination was normal. On trigeminal nerve testing, corneal reflex was absent on left side. Paresthesia was present over the left periorbital/forehead region, suggesting left sided trigeminal nerve involvement. Rest of the cranial nerve examination was found to be normal. Respiratory system, cardiovascular system, and per abdomen examination were essentially normal. Ear, nose, throat (ENT) consultation was done to look for any local cause and no abnormality was seen.

On investigations, her hemoglobin was 12.4 gm/dL, with total leucocyte count of 8500/cumm. Blood sugar was 92 mg/dL with normal liver and renal function tests. Her ESR was 07 mm in first hour and Mantoux test was negative. Human immunodeficiency virus testing by enzyme-linked immunosorbent assay was negative. ANA and ANCA testing was found to be negative. Cerebrospinal fluid (CSF) analysis was also done and no abnormality was found in CSF. ECG, X-ray chest, and ultrasound abdomen revealed no abnormal findings.

Radiological investigation in the form of MRI brain showed the lesion leading to expansion of left cavernous sinus (Figure 3). Rest of the differentials of cavernous sinus involvement were ruled out on the basis of history, physical examination, and relevant investigations. The patient was started with intravenous methylprednisolone for initial 3 days and oral corticosteroids thereafter. She responded very well to treatment

Table 1: Extraocular examination of left eye

Extraocular muscle of left eye	Function	Movement
Medial rectus	Adduction	Absent
Superior rectus	Elevator in abduction	Absent
Inferior rectus	Depressor in abduction	Absent
Inferior oblique	Elevator in adduction	Absent
Superior oblique	Depressor in adduction	Present but not full
Lateral rectus	Abduction	Absent

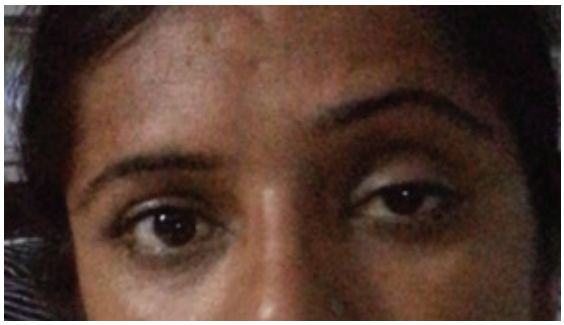

Figure 1: Left sided ptosis.

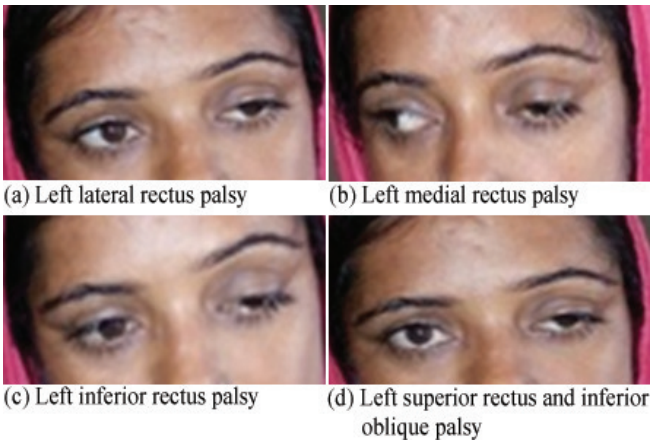

Figure 2: Restricted movements of left eye.

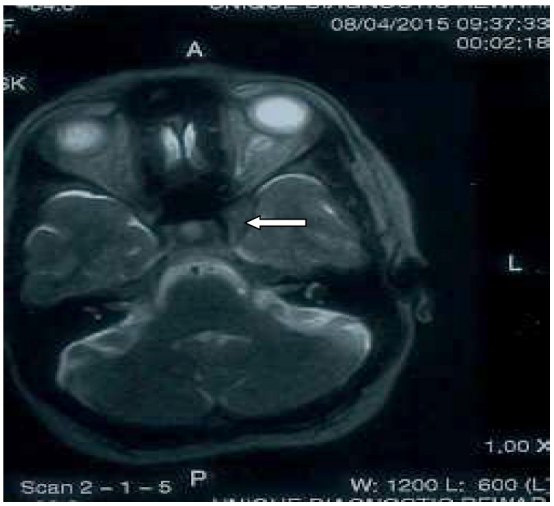

Figure 3: Brain MRI with T1W image showing a lesion in left cavernous sinus leading to its enlargement.

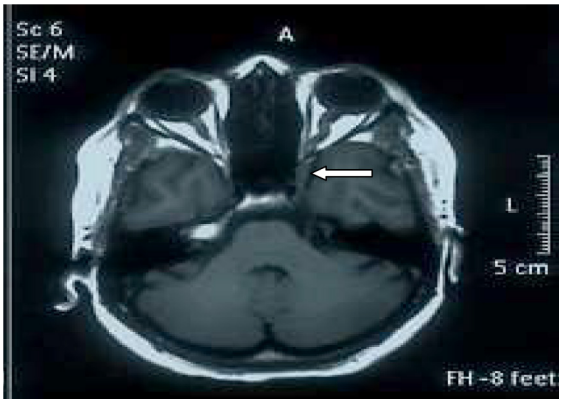

Figure 4: CEMR Brain with T1W image showing left cavernous sinus, reduced in size after 6 weeks of treatment.

with relief of headache 1 day after starting the corticosteroids. Improvement in ocular palsy was also observed during hospital stay and she was discharged from the hospital on oral corticosteroids. On follow-up she was pain free, with no ptosis or diplopia on left side. A follow up MRI scan of brain showed expansion of left cavernous sinus still present but the size was decreased compared to prior one (Figure 4).

On the basis of typical clinical features and MRI brain findings, a final diagnosis of THS was made.

Discussion

Although, the exact etiopathogenesis of THS largely remains unknown, it can be taken as a separate rare clinical entity. THS is caused by a non-specific inflammatory process in the cavernous sinus or superior orbital fissure (SOF). Pathologically, the inflammatory process is characterized by non-caseating, giant cell granuloma, fibroblast, lymphocyte, and plasma cell proliferation within the cavernous sinus septa and its walls.[2,3] The infiltration of the cavernous sinus with a non-specific inflammatory tissue leads to compressive neuropathy of the cranial nerves third, fourth, and sixth and V1 and V2 segments of trigeminal nerve, inside the cavernous sinus. The inflammatory process may also extend beyond the cavernous sinus/SOF, as is supported by involvement of other nerves such as optic and facial nerves in some case reports.[4-7]

Although, painful ophthalmoplegia is not rare, but THS as a cause of painful ophthalmoplegia have been considered as a rare clinical entity. The etiopathogenesis of THS largely remains unknown. No exact information is available on what actually triggers the acute inflammatory process within the cavernous sinus/superior oblique fissure. Thus, the syndrome can be taken as a manifestation of idiopathic orbital inflammation (Pseudotumor).[8] The initial patient evaluation with careful history and physical examination in a patient of painful ophthalmoplegia, localizes the lesion to cavernous sinus/superior oblique fissure. Thus, while working up a patient of painful ophthalmoplegia, the clinicians should be aware of the causes of parasellar syndrome and other causes of painful ophthalmoplegia. The causes of parasellar syndrome leading to painful ophthalmoplegia generally include trauma, neoplasms, inflammatory, and vascular pathologies. Contrast enhanced MRI with multiple views is the initial investigation of choice. MR imaging generally depicts an area of abnormal enhancement in the area of cavernous sinus in most of the cases of THS, but not in all the cases.[9-13]

Before the diagnosis of THS is made, one should rule out other various causes of painful ophthalmoplegia by a careful history, clinical examination, and the set of necessary investigations.

The International Headache Society (IHS) criteria for THS[14,15] include the following:

- Episode(s) of unilateral orbital pain for an average of 8 weeks if left untreated.
- Associated paresis of the third, fourth, or sixth cranial nerves, which may coincide with onset of pain or follow it by a period of up to 2 weeks.
- Pain that is relieved within 72 h of steroid therapy initiation.
- Exclusion of other conditions by neuroimaging and (not compulsory) angiography.

Even though the pathologic process and symptoms of THS are self-limiting, a short course of steroid is helpful in prompt relief and avoiding long-term complications. Steroids are the treatment of choice, which generally provide a relief from pain within 24 h to 72 h of starting the therapy.[16] Ophthalmoparesis usually requires a long course of steroid therapy ranging from weeks to months and even it may persist or resolve incompletely in some cases. In refractory cases or in patients not tolerating steroid therapy, other immunosuppressants such as azathioprine, methotrexate, or radiation therapy may be employed.[17]

Thus, the patient in our case report fulfilled all the four criteria led down by IHS for the diagnosis of THS. The patient presented with typical clinical features of unilateral headache/periorbital pain with associated involvement of the third, fourth, and sixth cranial nerves. Neuroimaging in the form of MRI brain ruled out other differentials. Also there was a prompt relief of pain to the patient, after start of the corticosteroid therapy.

Conclusion

THS is a diagnosis of exclusion and it must be carefully differentiated from more malignant causes of cavernous sinus involvement and painful ophthalmoplegia.

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