INTRODUCTION

Painful ophthalmoplegia is caused by a number of lesions in the parasellar, cavernous sinus and orbital apex regions. These include trauma to the skull base in this region, vascular abnormalities such as carotid-cavernous fistula and aneurysms, neoplasms such as pituitary adenoma, meningioma, lymphoma and metastasis, and inflammation around cavernous sinus and orbital apex which may induce cavernous sinus thrombosis.[1-3] Other causes include diabetic ophthalmoplegia, ophthalmoplegic migraine, sarcoidosis, eosinophilic granuloma, Wegner’s granulomatosis and orbital pseudotumor.[3,4]

Tolosa–Hunt syndrome (THS) was named after Tolosa and Hunt who separately described this condition as a granulomatous inflammation of the cavernous sinus and identified it to be steroid responsive.[1] They made the diagnosis after autopsy and surgical exploration. Modern imaging particularly magnetic resonance imaging (MRI) made it possible to study the lesions in the cavernous sinus and orbital apex responsible for this syndrome and other causes of painful ophthalmoplegia with high precision.[6]

CASE REPORT

A 55-year-old woman presented with the history of left hemicranial headache for the last 2 weeks followed by ptosis of the left eye upper eyelid 5 days before presentation. On examination, she was afebrile,
conscious and oriented. The pulse rate was 80 and the blood pressure was 130/80 mmHg. No signs of meningeal irritation were present; Left sided oculomotor, trochlear and abducens nerve palsies were present on evaluation of ocular muscles; pupil was spared. Sensory loss in the distribution of ophthalmic subdivision of trigeminal nerve was detected. There was no proptosis or chemosis; visual acuity was 20/20. Respective cranial nerves on the right side and the rest of the neurological examinations were normal. Biomicroscopy and funduscopy were unremarkable. Blood investigation including erythrocyte sedimentation rate, total leukocyte count, hemoglobin, and blood sugar were within normal range. Liver, thyroid and kidney function tests were normal. Serum for antinuclear antibodies was negative; lumbar puncture did not reveal any cerebrospinal fluid (CSF) abnormality. MRI of brain was performed with a 1.5 Tesla MR system applying T1-weighted (T1W) sequence before and after intravenous administration of gadolinium; T2W sequence was applied as routine 5 mm thick sequence in axial and coronal planes and as 1 mm thick axial constructive interference steady state (CISS) sequence. T2W sequence showed hypointense signal intensity lesion in the left cavernous sinus lateral to internal carotid artery [Figure 1]; the lesion was isointense on T1W sequence. Axial CISS image showed the left cavernous sinus lesion as a hypointense signal intensity mass extending towards orbital apex and superior orbital fissure [Figure 2]. Postcontrast T1W image showed homogenous enhancement of the lesion in cavernous sinus extending towards the orbital apex [Figures 3 and 4]. Patient was put on steroid therapy, initially intravenous methylprednisolone (160 mg) for 3 days followed by oral prednisolone (60 mg) for 8 weeks with gradual tapering. Patient reported significant relief of pain after 2 days; ophthalmoplegia responded gradually and resolved after 6 weeks when the neuro-ophthalmologic examination was normal. Repeat MRI was normal and consequently the diagnosis of THS was confirmed.

DISCUSSION

Tolosa–Hunt syndrome (THS) was named after Tolosa and Hunt who separately described this condition as a granulomatous inflammation of the cavernous sinus and identified it to be steroid responsive.[1] They made the diagnosis after autopsy and surgical exploration. Modern imaging particularly MRI made it possible to study the lesions in the cavernous sinus and orbital apex responsible for this syndrome and other causes of painful ophthalmoplegia with high precision.[2]

Painful ophthalmoplegia is caused by a number of lesions in the parasellar, cavernous sinus and orbital apex regions. These include trauma to the skull base in this region, vascular abnormalities such as carotid-cavernous fistula and aneurysms, neoplasms such as pituitary adenoma, meningioma, lymphoma and metastasis, and inflammation around cavernous sinus and orbital apex which may induce cavernous sinus thrombosis.[3] Other causes include diabetic ophthalmoplegia, ophthalmoplegic migraine, sarcoidosis, eosinophilic granuloma, Wegner’s granulomatosis and orbital pseudotumor.[3,4]

Tolosa–Hunt syndrome is considered as a nonspecific inflammation like orbital pseudotumor that results in painful ophthalmoplegia with a more posterior involvement in the region of orbital apex, superior orbital fissure and cavernous sinus.[5] It has been considered as a diagnosis of exclusion after excluding other known causes of painful ophthalmoplegia with proper clinical and laboratory evaluation. However, recent advancement in the imaging using computed...
Tolosa–Hunt Syndrome Demonstrated by CISS MRI; Wani et al

Imaging with MRI rules out other pathologies such as neoplasms and vascular abnormalities around the cavernous sinus as the cause of symptoms. MRI in THS shows enlargement of cavernous sinus with convex lateral wall due to soft tissue lesion within the sinus. The soft tissue lesion shows isointense signal intensity on T1W and iso- to hypointense signal intensity on T2W sequences with brisk enhancement on contrast administration. The follow-up MRI shows resolution of soft tissue lesion in the cavernous sinus which supports the diagnosis and also identifies the relapse early, if any. 

Constructive interference steady state is a high contrast and high spatial resolution MRI sequence that helps in differentiating small lesions in the cranial nerves within CSF filled spaces using T2W imaging. Close apposition of CSF spaces with the cavernous sinus and orbital apex makes demonstration of even small lesions within these structures evident, as hypointense signal void against the adjacent bright CSF. CISS images in our case depicted left cavernous sinus as enlarged and hypointense in signal intensity which extended towards the orbital apex. Conventional spin-echo T2W MRI in coronal plane did show the cavernous sinus lesion, but the bulk of the lesion and its true extent were not delineated by coronal and axial T2W MRI. Axial CISS MRI revealed the lesion and its extent completely. CISS appears to be more sensitive than conventional T2W MRI showing the true extent of the lesion as seen on contrast enhanced MRI. In the follow-up, resolution or relapse of the lesion may be shown with CISS with high sensitivity even without contrast administration.

THS has a tendency to recur and if untreated, may result in visual loss and other morbidities. Treatment is performed with steroids. Dramatic response to steroid administration with prompt relief of pain and slow resolution of ophthalmoplegia confirms the diagnosis of THS differentiating it from other causes of painful ophthalmoplegia. Repeat MRI further supports the diagnosis by demonstrating resolution of the inflammatory lesion. Neurosurgical procedure for biopsy is rarely necessary.

We conclude that THS is a benign, steroid responsive nonspecific granulomatous inflammation of the cavernous sinus that should be suspected in a case of painful ophthalmoplegia. MRI not only excludes other causes but in fact, may demonstrate the offending inflammatory lesion itself. CISS MRI imaging can demonstrate even slight degree of enlargement of the cavernous sinus in THS.

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Conflicts of Interest
There are no conflicts of interest.

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