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Case Report

Tolosa Hunt syndrome: a rare syndrome

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ABSTRACT

Tolosa Hunt Syndrome (THS) is rare syndrome with an estimated annual incidence of 1 case per million per year. THS is painful ophthalmoplegia caused by nonspecific inflammation of cavernous sinus or superior orbital fissure. It is often unilateral with severe headache and ophthalmoplegia involving third, fourth, fifth and sixth cranial nerves. We present a case admitted in our hospital that came with complaint of severe unilateral headache with loss of vision of left eye. He did not have any other significant history except that he was complaining of partial sensory loss over left upper part of face. Ophthalmologist reference was taken to rule out any other cause involving optic disc and funduscopy which was normal. Further investigations were done which ruled out all possible causes. Patient was started on steroids and on MRI scan and clinical presentation, patient was diagnosed as Tolosa- Hunt Syndrome. Thus we report a rare case of THS which showed gradual recovery with corticosteroids.

Keywords: Tolosa Hunt Syndrome, Granulomatous inflammation, Cavernous sinus, Superior orbital fissure

INTRODUCTION

Tolosa Hunt syndrome (THS) was initially described by E Tolosa in 1954 and then by W. E Hunt et al, in 1961. The condition was termed Tolosa-Hunt syndrome by Smith and Taxdal in 1966. An estimated annual incidence of THS is 1 case per million per year. Males and females are equally affected.[1,2]

THS is uncommon disorder causing nonspecific granulomatous inflammation of the septa and wall of the cavernous sinus, with a lymphocyte and plasma cell infiltration, giant cell granulomas, and proliferation of fibroblasts.[3] It is characterized by severe and unilateral headaches with extra ocular palsies, usually involving the third, fourth, fifth, and sixth cranial nerves, and ophthalmoplegia. It is most often unilateral. The aetiology is unknown, although it shares histopathological features with idiopathic orbital pseudo tumour. Clinically, painful ophthalmoplegia or ophthalmoparesis and immediate response to steroid therapy are a hallmark of the condition.

THS diagnostic criteria were provided by the International Headache Society (IHS).

Modified revised IHS headache classification of 2004:

Description

Episodic orbital pain associated with paralysis of one or more of the third, fourth and/or sixth cranial nerves which usually resolves spontaneously but tends to relapse and remit.

Diagnostic criteria

A. One or more episodes of unilateral orbital pain persisting for weeks if untreated.
B. Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or demonstration of granulomas by MRI or biopsy.
C. Paresis coincides with the onset of pain or follows it within 2 weeks.
D. Pain and paresis resolve within 72 h when treated adequately with corticosteroids.

E. Other causes have been excluded by appropriate investigations.

Some reported cases of Tolosa-Hunt syndrome had additional involvement of the trigeminal nerve (commonly the first; division) or optic, facial or acoustic nerves. Sympathetic innervation of the pupil is occasionally affected.

The syndrome has been caused by granulomatous material in the cavernous sinus, superior orbital fissure or orbit in some biopsied cases.

Careful follow-up is required to exclude other possible causes of painful ophthalmoplegia.[4]

The clinical differential diagnosis of steroid responsive painful ophthalmoplegia includes metastases, carotid-cavernous fistulae, pituitary adenomas, vasculopathic cranial neuropathy, aspergillus invasion, Wegener's granulomatosis, sarcoidosis, lymphoma and ophthalmoplegic migraine.[5]

CASE REPORT

A 60 year old male presented with history of unilateral headache on left side with painful left eye since 8 days. Followed by which he had history of progressive diminision of vision of left eye which was gradual in onset over 7 days. He also complained of decrease in eye movement of left side. He was a known case of diabetes mellitus type II on treatment since 2 year and operated for cataract surgery both eyes.

On clinical examination, his vitals were stable. He had left sided ptosis with left lateral rectus palsy. He had difficulty with left medial, up and down eye movements (Figure 1). Decrease visual acuity of left eye suggestive of optic (II^{nd}) nerve involvement. He also had left occulomotor (III^{rd}) nerve palsy with left abducens (VI^{th}) nerve palsy (Figure 2). Patient also had loss of sensation of upper part of left side of face which was significant of involvement of ophthalmic branch of trigeminal nerve (V^{th} nerve). No any other significant systemic finding was present. Ophthalmologist opinion was taken to rule out any other cause of headache and loss of vision. Funduscopy was normal.

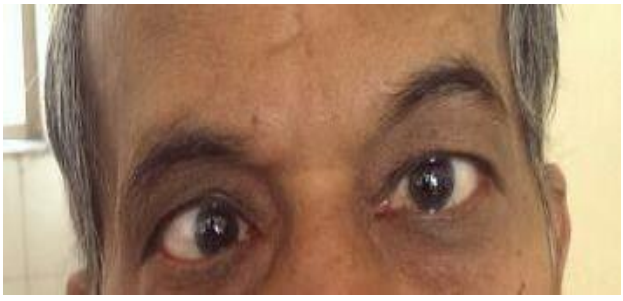

Figure 1: Difficulty in left medial up and down eye movements.

Blood investigations were normal with good control of sugars. Lumbar puncture was nonspecific. Rest all blood reports were insignificant.

Figure 2: 3rd and 6th nerve palsy.

He was advised MRI brain which was suggestive of enhancing mass lesion in the orbital apex extending into the left superior orbital fissure.

CT brain with contrast was done which was suggestive of moderately enhancing soft tissue in the left cavernous sinus extending anteriorly through the superior orbital fissure into the left orbital apex encasing the cavernous portion of the left internal carotid artery and the left optic nerve (Figure 3).

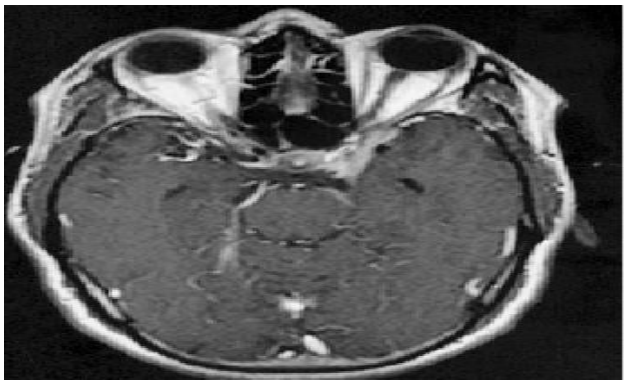

Figure 3: CT scan showing enhancing soft tissue in left cavernous sinus.

The patient was thus started on intravenous methylprednisolone followed by oral steroids after 3 injectable doses of steroids. He showed a progressive recovery within 48 hours with complete relief from symptoms. And there was no recurrence on follow-up. Biopsy couldn't be done because he and his relatives were not willing for same.

DISCUSSION

Tolosa Hunt Syndrome is a rare disorder caused by nonspecific granulomatous inflammation of cavernous sinus or superior orbital fissure. There is no other systemic involvement. Recurrence is possible. Prognosis with help of steroid shows good recovery. Other differential diagnosis has to be ruled out like sarcoidosis,

meningioma, lymphoma, thrombosis of cavernous sinus and others. As its diagnosis of exclusion, thus MRI and CT scan is essential part in evaluating such patient with painful ophthalmoplegia. And also it is important to rule out any ophthalmic cause causing headache, pain and loss of vision.

Thus with the discussion of the above case, it is distinctive neuroimaging and response to steroids with resolution which differentiates THS with other possibilities.

CONCLUSION

Early diagnosis with the help of neuroimaging techniques and its clinical correlation is essential for early management thus improving outcome.

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REFERENCES

1. Sathyanathan BP, Rajasundaram R, Sankaravadivelu ST, Nadhamuni K. A case of tolosa-hunt syndrome - Mr imaging appearance Neuroradiology. 2006;16(1).
2. Kline LB, Hoyt WF. The Tolosa-Hunt syndrome J Neurol Neurosurg Psychiatry. 2001;71:577-82.
3. Barnard B, Hurter D, Roux F. Tolosa -Hunt syndrome. SA Journal of Radiology, 2012.
4. Lance JW, Olesen J. The International Classification of Headache Disorders ICHD-II. Cephalalgia, 2004.
5. Khera PS, Singh S, Chowdhury V. Tolosa Hunt Syndrome: A Case Report. Ind J RadiolImag. 2006;16(2):175-7.

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