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CASE SERIES OF A RARE DISEASE: TOLOSA HUNT SYNDROME

General Medicine

Dr. Maitri Mehta* Junior Resident in Department of General Medicine, Dr. D. Y Patil School of Medicine, Nerul, Navi Mumbai. *Corresponding Author

Dr. Sonalika Dughar Junior Resident in Department of General Medicine, Dr. D. Y Patil School of Medicine, Nerul, Navi Mumbai.

Dr. Nivedita Moulick Guide and HOD- Department of General Medicine, Dr. D. Y Patil School of Medicine, Nerul, Navi Mumbai.

ABSTRACT

Tolosa Hunt syndrome is a non-specific inflammation of cavernous sinus or superior orbital fissure causing painful ophthalmoplegia. It is a rare disease with limited number of cases reported till date. It was first described by Tolosa in 1954 and seven years later was explored further by Hunt. The estimated annual incidence is one case per million per year. Characteristic clinical manifestations include recurrent retro-orbital pain, with palsies of third, fourth or sixth cranial nerves. We sought to present case series of 4 patients of Tolosa Hunt syndrome.

KEYWORDS

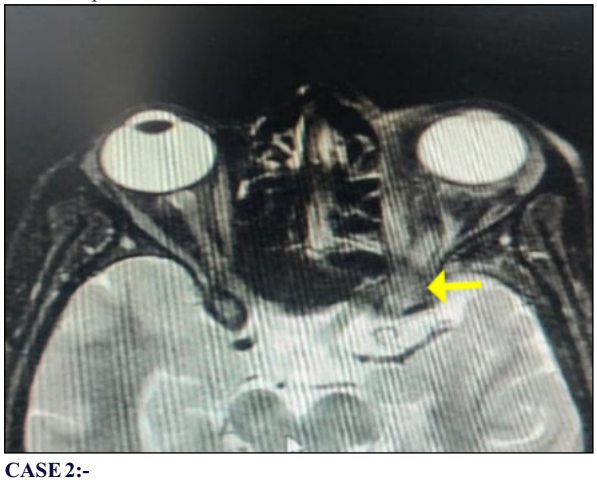
INTRODUCTION:-

Tolosa-Hunt syndrome (THS) is an uncommon idiopathic non-specific inflammatory disease of the cavernous sinus, superior orbital fissure, or orbit associated with ocular pain and ophthalmoplegia. It affects males and females equally. The average age of onset is 41 years, but there have been cases reported among people younger than age 30. In the presence of painful ophthalmoplegia, the finding by MRI of cavernous sinus enlargement, with the herein described signal and extension characteristics and slow resolution with corticosteroid treatment, is highly suggestive of the Tolosa-Hunt syndrome.

There are relatively few reports on the clinical course of THS, so the objective of this retrospective study was to investigate the clinical outcome.

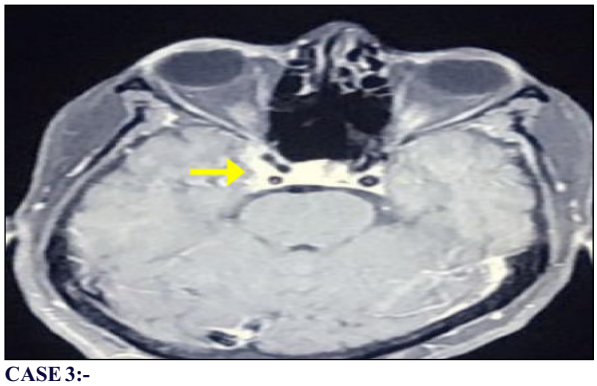
CASE 1:-

A, 45 year old, female came with complains of left sided headache since 1 month, left eye pain since 10 days, drooping of eyelid and diplopia since 10 days. No history of comorbidities. On examination of left eye, there was complete ptosis, no extraocular movement, pupils were 3mm in size and sluggishly reacting to light and no colour vision. However right eye examination findings was normal. Fundus examination of both eyes was normal. MRI brain+ bilateral orbit was suggestive of minimal relative prominence of perioptic space on left side as compared to right side. CSF study was normal. Patient was started on steroids intravenously. Patient responded well to steroids. Patient responded well to steroids within 36 hours.



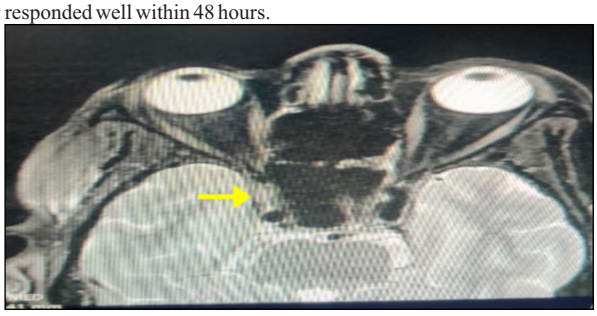
CASE 2:-

A, 40 year old, female, came with complains of right sided headache since 1 week, drooping of right eyelid and lag ophthalmos since 1 week. The patient was diagnosed diabetes mellitus since 1 month. On examination of right eye, there was ptosis of right eye, no extraocular movement, pupil was 3mm in size normally reacting to light and colour vision was intact. Left eye examination findings were normal. Fundus examination of both eyes was normal. MRI brain+ orbit was suggestive of enhancing Dural based soft tissue lesions in medial aspect and right middle cerebellar fossa suggestive of inflammatory pseudotumour. CSF study was done to rule out idiopathic pachymeningitis which was normal. Patient was started on steroids intravenously. Patient responded well to steroids and responded well within 48 hours.



CASE 3:-

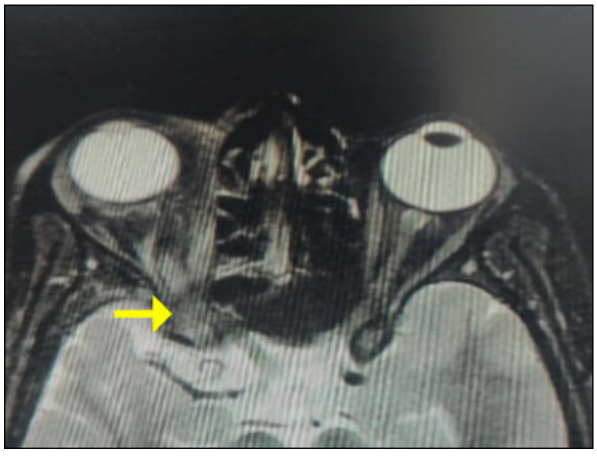
A, 63 year old, female came with complains of redness and pain in right eye since 1 month, Diminision of vision in right eye, lag ophthalmos and deviation of angle of mouth to left side since 1 month. Patient had similar episode 9 years back during which MRI brain + orbit was suggestive of enhancing Dural based soft tissue lesions in medial aspect and right middle cerebellar fossa suggestive of inflammatory pseudotumour. Patient was then started on steroids after which the symptoms resolved. On examination of right eye, there was ptosis , no extraocular movements ,pupil was 3mm normally reacting to light and colour vision was intact. Left eye examination findings were normal. Fundus examination of both eyes was normal. MRI brain+ orbit was suggestive of right perioptic space prominence. CSF study was normal. Patient was started on steroids intravenously. Patient responded well within 48 hours.



CASE 4:-

A, 61 year old, female, came with complains of right sided headache and right eye pain since 5 days and diplopia since 5 days. The patient is known case of diabetes mellitus, hypertension and ischaemic heart disease. On examination of right eye, there was ptosis, no extraocular movement, pupil was 3mm normally reacting to light and colour vision was intact. Left eye examination findings were normal. Fundus examination of both eyes was normal. MRI brain+ orbit was suggestive of minimal relative prominence of perioptic space on right side as compared to left side. CSF study was normal.

Patient was started on steroids intravenously. Patient responded well with steroids within 24 hours.



DISCUSSION:-

The diagnosis of Tolosa-Hunt syndrome is suspected based upon the presence of characteristic physical features (e.g., pain, headache, ophthalmoplegia). The diagnosis may be confirmed by a thorough clinical evaluation, detailed patient history, radiologic tests including computed tomography (CT) scan, and magnetic resonance imaging (MRI). Corticosteroids are the treatment of choice for Tolosa-Hunt syndrome (THS), usually providing significant pain relief within 24–72 hours of therapy initiation. Ophthalmoparesis usually resolves within weeks or months. Ophthalmoparesis may not resolve completely in some cases depending on the degree of inflammation and the aggressiveness of therapy. For refractory cases, azathioprine (Imuran), methotrexate, or radiation therapy has been employed. The prognosis for Tolosa-Hunt syndrome is good. Spontaneous remissions can occur; relapses may occur in up to 40% of the patients.

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