Tolosa–Hunt syndrome

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Abstract

Aim: This study is aimed to report Tolosa-Hunt Syndrome a rare case.

Introduction: A non inflammatory disorder in cavournous sinus or in the fissure of orbit leading to painful eye movements is called as Tolosa Hunt syndrome. **Case report:** A young married house wife, with history of worsening eye pain, double vision, decreased sensation on the right side of face, and ptosis was referred by her local practitioner to the Neurology Department of Peoples Medical University Hospital Nawabshah, Sindh, Pakistan, in the month of October 2018. She was admitted, detailed history, general physical and systemic examination and investigations were carried out. Keeping in the view of history, examination and investigations THS diagnosis was made. Prompt treatment was started; her clinical symptoms were alleviated to a great extent within three days of admission and she was discharged well to home after 4 days of admission. **Conclusion:** Tolosa hunt syndrome is very rare disease; it is diagnosis of exclusion, when all other diseases with same clinical features of TSH diagnosis were excluded. Early diagnosis and prompt treatment improves the prognosis and out comes in this condition.

Key words: Tolosa-Hunt Syndrome, Diplopia, Ophthalmoplegia.

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INTRODUCTION:

THS (Tolosa Hunt syndrome) is a distracted inflammatory changes of any cavernous sinus & orbital fissure secondary to hurting ophthalmoplegia [1].

THS is non-fatal ailment and is characterized by severe one sided headache associated with orbital pain and weakness and paralysis of certain extra

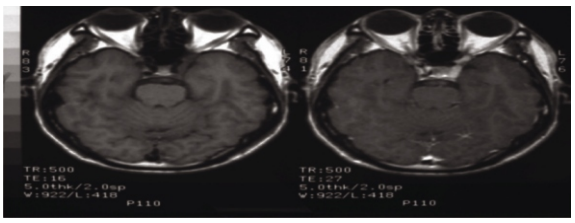
ocular muscles leading to ophthalmoplegia and extra ocular palsies [1].International Society for Headache in 2004 delivered the criterion for the definition and diagnosis of THS which also comprised the granuloma [2]. It is a rare disease in United States as well as globally. Tolosa hunt syndrome occurs less likely in the first two decades of life. It seems to have an un fluctuating dissemination in subjects over the age of 20 years. When children are affected by tolosa hunt syndrome, the course of the disease looks to be analogous to the adults [3]. Both genders are affected likewise, and it is important to put this syndrome in differential diagnosis in the children [4].

History:

A young female presented in emergency department with history of severer retro orbital

pain on right side of face and diplopia. She also complains of visual loss and facial paresthesia for 10 days. **On examination:** There were painful eye movements with ptosis on right side. There was no lid edema. Mild proptosis with optic disc edema was noted. The ipsilateral loss of corneal reflex was noted.

Investigation: All the routine investigations were performed including the CBC, ESR 12mm/hr, serum urea 35mg/dl, sugar 110mg%, creatinine 1.1mg/dl, electrolytes HIV negative. Thyroid function test T3, T4 and TSH were normal ranges, FTA (fluorescent treponeal antibody test), ANA (Antinuclear antibody test), ANCA (Anti-neutrophill cytoplasm antibody) test was negative. Gram staining was negative, bacterial cultures shown no growth, cytology was negative, and opening pressure of CSF was carried out and was in normal range. The above all investigations were within normal limits. In plain and contrast CT scan brain and orbit imaging, in cavernous sinus and **in the fissure of orbit** there were changes with inflammation on right side. There was enhancement of the cavernous sinus on MRI (plain and contrast) brain and orbit on the right side. Cranial nerve examination had shown involvement of 3rd , 4th , 5th (ophthalmic division) and 6th cranial nerve were involved on right side.



After confirming the judgment of TSH treatment with injectable corticosteroids was initiated, a significant reduction in pain was claimed by patient within 24 hours of treatment start and the pain was completely subsided in about 72 hours of initiating the steroid therapy.

Discussion:

In the majority subjectsof THS the exaggerated subjects practice extreme pointed tenderness & weakness of musculature tissues around the eye, the features are commonly restricted to one side of face [5]. Decrease in symptoms can occur without any medical interference, and can reappear without any clear character [6].

In current case scenario a young female presented with severe retro orbital pain, diplopia, and facial paresthesia on right side of face, with painful eye movements, ptosis and ipsilateral loss of corneal reflex on examination. Large number of investigations performed, inflammatory changes were noted on radiology, treated with corticosteroids with excellent improvement in clinical features. All the features of presentation, examination, investigations done in this case are similar to that of the other reported cases.

The diseased subjects may have weakness of different CN, and flaccid superior lid of eye leading to drooping. Fever, chronic tiredness, unsteadiness, arthralgia and double vision are the clinical features in subjects of THS. Sometimes patient may complain with perception of protrusion of eyeball (one or both) [5,6].

Ptosis and retro orbital pain both were also experienced by the subject in our case. No particular cause of TSH was recognized yet. The inflammation of regions behind the eyes that is the cavernous sinus thrombosis and superior orbital fissure were assumed & related with THS. No obvious cause was detected in present case.TSH is commonly verdict of keeping out; an extensive range of laboratory investigations is required to rule out other causes, such as CBC, thyroid profile, serum proteins electrophoresis, CSF analysis and many others [5].We also made the THS

diagnosis on available statistics and investigation.

The inflammatory change in cavernous sinus, orbital apex & superior orbital fissure could be detected by plan and/or contrast Computed Tomographic scan, MRI orbit & brain. MRA or digital subtraction of brain and orbit are also useful tools in diagnosis of THS [5].

On cross sectional imaging the inflammatory alteration of orbit & absenteeism of CN palsy usually rules out THS, & this favors the diagnosis of a more benign condition called orbital pseudo tumor. Occasionally biopsy may be needed to indorse the diagnosis and rule out the neoplasm [5]. Craniopharyngioma, migraine and meningioma are considered in differential diagnosis of THS [5].The above diagnoses was not suggested here on the basis of history, examination and investigations.

Immune suppressive drug prednisolone is preferred from other corticosteroids to treat this syndrome, or steroid sparing drugs as azathioprine or methotrexate are also included in the treatment of THS [5].We had treated this case with injectable steroids and got improvement a lot here. Radiotherapy also had been suggested [7]. THS has usually good prognosis. Subjects mostly response to the corticosteroids, remissions may be seen spontaneously, though the damage to the movement of ocular muscles may persist [5]. Relapse occurs in round about 30% to 40% of THS treated subjects [5]. From New Zealand and New South Wales Australia only single case from each has been reported [5].

THS is a rare condition in Pakistan; it can affect any age but usually occurs after second decade of life, the reported case is a young age female.

Conclusion

Tolosa hunt syndrome is very rare disease; it is diagnosis to exclude the other diseases when no evidence of other disease is there. Early diagnosis and prompt treatment improves the prognosis and outcome of this condition.

References

1. La Mantia L, Curone M, Rapoport AM, Bussone G (2006). "Tolosa–Hunt syndrome: critical literature review based on IHS 2004 criteria". Cephalalgia. **26** (7): 772–81. doi:10.1111/j.1468-2982.2006.01115.x. PMID 16776691. Archived from the original on 2013-01-18.
2. "Tolosa–Hunt syndrome". Who Named It. Retrieved 2008-01-21.
3. 2Zanus C, Furlan C, Costa P, Cosentini D, Carrozzi M. The Tolosa-Hunt syndrome in children: a case report. Cephalalgia. 2009. 29:1232-1237
4. 32Pérez CA, Evangelista M. Evaluation and Management of Tolosa-Hunt Syndrome in Children: A Clinical Update. Pediatr Neurol. 2016 Sep. 62:18-26. [Medline].
5. Danette C Taylor, DO. "Tolosa–Hunt syndrome". eMedicine. Retrieved 2008-01-21.
6. "Tolosa Hunt Syndrome". National Organization for Rare Disorders, Inc. Retrieved 2008-01-21.
7. Foubert-Samier A, Sibon I, Maire JP, Tison F (2005). "Long-term cure of Tolosa–Hunt syndrome after low-dose focal radiotherapy". Headache. **45** (4): 389–91. doi:10.1111/j.1526-4610.2005.05077_5.x. PMID 15836581. Archived from the original on 2013-01-10.