

Tolosa Hunt Syndrome: A Case Report

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ABSTRACT

Tolosa Hunt Syndrome is a rare disorder characterized by severe and unilateral headache associated with painful and restricted eye movements. It is mainly due to paresis of one or more of the oculomotor (3rd), trochlear (4th), abducent(6th) cranial nerves caused by a granulomatous inflammation in the cavernous sinus, superior orbital fissure or orbit. In this case, a 61 year old male patient with complaints of headache, right complete ptosis with throbbing pain around right eye. This case report study has been presented for the consideration of steroid therapy in Tolosa Hunt Syndrome.

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INTRODUCTION

Tolosa Hunt Syndrome is a rare disorder, defined as painful ophthalmoplegia consists of periorbital/hemicranial pain associated with ipsilateral ocular motor nerve palsies, oculosympathetic paralysis and sensory loss in the distribution of the ophthalmic branch of trigeminal nerve and occasionally the maxillary division of the trigeminal nerve. Various combinations of these cranial nerve palsies may occur. There is a specific subset of patients who develop painful ophthalmoplegia due to a non-specific inflammatory process in the region of the cavernous sinus/ superior orbital fissure.

It is a steroid-responsive painful ophthalmoplegia which is secondary to idiopathic granulomatous inflammation. It has been categorized as a diagnosis of exclusion because of its non-specific radiologic presentation. The orbital apex pseudotumor and granulomatous inflammation of the cavernous sinus have same clinical features and should be considered as a part of the spectrum of Tolosa Hunt Syndrome. Symptomatic improvement after steroid therapy is an essential but is not an exact proof of the syndrome, because lesions such as lymphomas may also respond to steroids.

DIAGNOSIS

It is based on;

- Eye pain on one side of the head that persists for at least eight weeks if untreated.
- Associated damage to third-fourth/ sixth cranial nerves.
- Relief of pain within 48hrs after the administration of steroids.

Diagnosis may be confirmed by a thorough clinical evaluation

- Detailed patient history
- Radiologic tests like computed tomography (CT) scan and magnetic imaging (MRI)
- These examinations may show characteristic enlargement/ inflammation of the areas (cavernous sinus and superior orbital fissure).

CASE REPORT

A 61 year old male patient came with complaints of headache, right ptosis, pain over right eye for 2 days. He had woken up in the middle of the night with severe headache bifrontally which is throbbing in nature, and also had woken up in the next morning with partial ptosis of right eye. By evening it progressed to right complete ptosis with throbbing pain around right eye.

On examination 3rd cranial nerve ophthalmic nerve (V1) impairment noticed but there was no facial palsy. MRI showed normal cavernous sinus region, right superior rectus and levator palpebrae superioris complex was thickened and had minimal contrast uptake. CSF tapping was done under aseptic precaution. In which, CSF protein and glucose was found to be elevated. Patient showed positive bitot's spot and left conjunctival congestion. He had k/c/o type II DM for 10 year, hemorrhoids for 10 years, 4 years back a blood transfusion was done for severe anemia and also had a history of acute bronchitis 2 weeks ago. Initially patient was given T. Deflazacort 6 mg but patient didn't show any improvement. The selective involvements of muscles

generated discussion towards a possible neuromuscular junction disorder. Repetitive nerve stimulation and neostigmine test were performed and results were negative. After that methylprednisolone pulse therapy was given, then patient showed symptomatic improvement. For pain T. ultracet was given. On discharge inj. Methylprednisolone was changed to T. wysolone 40 mg for 2 weeks.

DISCUSSION

It is usually affects only one eye (unilateral). Affected individuals experience intense sharp pain and decreased eye movements. Symptoms often will subside without spontaneous remission and may reoccur randomly. These individuals may exhibit signs of paralysis of certain cranial nerves such as drooping of the upper eyelid (ptosis), double vision (diplopia), large pupil and facial numbness. The affected eye may also exhibit protrusion of the eye (proptosis). The main cause of Tolosa Hunt Syndrome is unknown, but the disorder is thought to be associated with inflammation of specific areas behind the eye like cavernous sinus and superior orbital fissure. The average age of onset is 41 yrs, but it had been reported among people younger than 30yrs. Methylprednisolone and prednisolone are found to be effective in Tolosa Hunt Syndrome. MRI findings before and after corticosteroid therapy are the important diagnostic criteria to differentiate it from other cavernous sinus lesions that stimulate THS both clinically and radiologically.

CONCLUSION

The pain associated with Tolosa Hunt Syndrome subsides with the use of short term use of steroid drugs. Pain is usually reduced in untreated cases within fifteen to twenty days. With steroid treatment, pain typically briskly subsides within twenty four to seventy two hours. The diagnosis is

based on the brisk steroid response. Although steroids are generally tapered over weeks to months, in some cases prolonged therapy may be necessary. The affected individuals may be vulnerable to recurrent future attacks. Steroids like prednisone and methylprednisolone are effective in Tolosa Hunt Syndrome than other steroids. Immunosuppressive agents like azathioprine and methotrexate can be beneficial if patients show no response to steroids.

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