Review Article

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Tolosa-hunt syndrome: a 48year male with history of facial palsy

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Abstract

Tolosa-Hunt syndrome is a rare disease characterized by intense periorbital headaches, along with ophthalmoplegia. Symptoms are usually unilateral. A 48-year-old male presented with right eye dropping (R ptosis) and double vision from 2 weeks, which was progressive along with pain involving LMN VII palsy. The pain was severe in intensity, continuous and sharp. It had been diagnosed as Tolosa-Hunt syndrome based on MRI, steroid response, CSF analysis, other rule outing investigations. Corticosteroids were prescribed as the treatment. The patient found to be better with the therapy and tapering was done. Later, due to fontal headache azathioprine added as immunosuppressants. The clinical symptoms are similar to other diseases like migraine headache, orbital cellulitis. This case report focus on the diagnosis and treatment of Tolosa hunt syndrome.

Keywords: Tolosa-Hunt syndrome, Ophthalmoplegia, palsy.

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Introduction

Tolosa-Hunt syndrome is a rare disease characterized by intense periorbital headaches, along with ophthalmoplegia. Symptoms are usually unilateral.[1] The diagnosis of Tolosa-hunt is difficult because of the similarity of symptoms with other disorders. Therefore, timely and appropriate techniques (MRI) should perform along with relevant laboratory investigation for accurate diagnosis. Spontaneous remission of symptoms can occur without intervention and recurrence is without any pattern.[2] Major signs of this disorder ptosis, double vision, large pupil, and facial numbness. The condition should treat with steroids and response to steroid therapy is an identification mark of the condition.[3]

Here we present the case of a patient with painful ophthalmoplegia of the right eye for the past 2 weeks, its diagnostic workup, treatment and follow-up.

CASE PRESENTATION

A 48-year-old male presented with right eye dropping (R ptosis) and double vision from 2 weeks, which was progressive along with pain involving LMN VII palsy. The pain was severe in intensity, continuous and sharp. There was a history of facial palsy 8 years back and diplopia which improved with steroids. Also, he had a migraine 1 year back No history of hyperthermia, facial pain or hearing loss on the same side during the current hospital presentation. He had a weight of 74 kg and a height of 173 cm. On examination vitals (Intake/output, pulse rate, oxygen saturation, blood pressure, respiratory rate) found to be normal. The patient was conscious, oriented and external complete ophthalmoplegia identified. Based on initial symptoms provisionally diagnosed as ocular myotonia. The patient admitted to the hospital for a short time. During this time the vitals and input/output level found to be normal. The Glasgow scale showed E4V5M6 throughout the admission.

RNS: right orbital oculi CMAP decreased with a decremental response on RNS.

MRI brain with orbit found to be normal.

MRI brain: post-contrast enhances smooth meningeal thickening in right middle cranial fossa medially involving right cavernous sinus extending anteriorly up to right superior orbital fissure. Possibility of granulomatous meningitis.

During admission haemoglobin-15.6, WBC total- 10690, DC, Platelet count, PTT, INR, Urea, Creatinine, bilirubin, SGPT, SGOT, ALP, Serum protein total, albumin, globulin, sodium, potassium, calcium, cholesterol, magnesium found to be normal. The FBS and RBS level was fluctuating in this period.

ANA: Normal, ANA profile: Normal, CSF ACE level:0.20u/l

CSF analysis: TC-5cells, DC: Only lymph, sugar-122, protein-87.4mg%, fluid RBC 0-1 ADA-3, VDRL—negative

Grain stain- nil, fungal smear-nil, cryptococcus-nil, Serum ACE: 26.8 nmol/ml/min

Based on the subjective and objective data it was diagnosed as Tolosa-hunt syndrome. The patient managed with IV steroids and other supportive measures for 3 days. A patient prescribed with inj. Dexamethasone 2mg IV Q6H for 2 days and stopped, Inj Rantac (ranitidine) 150mmg IV Q12H, Inj. Iveprid (methylprednisolone) 1gm IV OD after discontinuation of Dexamethasone till discharge (3 days). Dermatological findings of extensive tinea versicolor face, back and neck identified and prescribed with T. AF (Fluconazole) 400 twice-weekly (Monday and Thursday) for 4 weeks, Anaboom AD lotion 0-0-1 for 4 weeks and KZ (ketoconazole)soap.

Condition of the patient improved (Steroid response), hence being discharged. Discharged with Tab. Predmet (methylprednisolone) 8mg 1-0-1 for 1week, then ½-0-1 for next 1 week, Tab. Control (pantoprazole) 40 mg 1-0-1 for 2 weeks, Tab. shelcal (Vitamin D3 and Calcium)500mg 0-1-0 for 2 weeks.

After 2 weeks the patient was followed up and given medicines for the next 4 weeks with T. Rantac 150mg 1-0-1, T. shelcal CT 0-1-0 and for 2 weeks T. Wysolone (prednisolone) 20mg -0-10 mg and for the next 2 weeks 20mg-0-0 were prescribed. FBS found to be elevated and prescribed with T. Gemer (glimepiride + metformin) 0.5gm 1-0-0 for 4 weeks. T.Gemer discontinued due to FBS normalisation After 4 weeks steroid reduced to 10mg OD followed by 5 mg after 1month. After 6 weeks apart from other drugs Tab. Azoran (azathioprine)50g ½-0-0 for 7days then 1-0-0 for 3 weeks and 1-0-1/2 for 4 weeks were added with therapy. On the next visit frontal headache complained by the patient therefore Wysolone increased to 10mg as OD with other drugs. After 10 days Tab. Azoran stopped and wysolone frequency increased to 1-0-1/2. The patient continued the treatment with steroid.

DISCUSSION

National Organisation for Rare Disorders (NORD) included Tolosa hunt syndrome as rare disorder. Frist patient reported on 1954 with left orbital pain, ipsilateral progressive visual loss, total left ophthalmoplegia and reduced sensation over the first division of the trigeminal nerve. Later, in 1961 six more patients identified with this syndrome. The cranial nerves involved were III, IV, VI and VI1.[4] In this case patient has LMN VII palsy with right eye dropping and diplopia.

The etiologies or exact cause of disorder is not known. But some theories are proposed based on abnormal autoimmune responses and inflammation mainly behind the eyes. There is no gender predominance in prevalence. The majority of patients are above 41 but there are paediatric or young patients. The differential diagnosis is important in Tolosa-hunt syndrome because the symptoms have similarities with orbital cellulitis, cavernous sinus thrombosis and migraine headaches. It can be differentiated using proper imaging and laboratories tests.

International Headache society have criteria for diagnosis of the disorder. Mainly unilateral eye pain that persists at least eight weeks if not treated, damage or irritation of cranial nerves (III, IV, V), steroid response, others test that rule out infection aneurysm.[4-5] In this case MRI, clinical signs, steroid response, other laboratory tests like CSF culture, CSF analysis, Serum ACE etc were tested for diagnosis. Also, the patient shown steroid response within 3 days. The primary treatment is steroid which reduce the symptoms and pain. The untreated condition will lead to reduction of pain within 3 – 4 weeks but recurrence chances is high.[6-8] If even treated, the recurrence of the same in future is not preventable. Studies are undergoing regarding the additional treatments like radiation therapy, immunosuppressive drugs. In this case azathioprine is prescribed as immunosuppressant for disease based on other studies.[5] Both corticosteroid and immunosuppressant are effective in these cases.

CONFLICT OF INTEREST: Nil

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