Recurrence of Tolosa-Hunt Syndrome Isolated to the Sixth Cranial Nerve

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Tolosa-Hunt syndrome (THS), first reported in 1954, is characterized by hemicranial headache, retrobulbar pain, with 1 or multiple cranial nerve palsies.[1] These clinical features are attributed to granulomatous inflammation of the cavernous sinus. Although THS is a diagnosis of exclusion, it is confirmed by observing cavernous sinus inflammation on magnetic resonance imaging (MRI). The treatment of choice is steroids followed by steroid-sparing agents.[2] Recurrence is reported in 50% of cases, usually on the ipsilateral side and rarely involving the contralateral side or bilaterally.[3] We wish to add a unique presentation of ipsilateral recurrence of THS involving the sixth cranial nerve to the limited literature on the subject.

Case Report

A 64-year-old man presented with complaints of diplopia and unilateral headache on the left side for 2 months. The diplopia was sudden in onset and worsened on movement of eyes to the left side. The headache involved the periorbital and retro-orbital region and had a dull and throbbing character. There were no associated neurological symptoms, and his history was insignificant.

On neurological and ophthalmologic examination, palsy of the left lateral rectus muscle was observed, indicating involvement of the left abducens nerve. Examination of the other cranial nerves revealed normal findings. Routine investigations revealed elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Cerebrospinal fluid analysis was normal. Cavernous sinus inflammation was observed on MRI, and a diagnosis of THS was established. The patient was prescribed prednisolone with a tapering regimen for 4 weeks, leading to resolution of symptoms.

After 2 months of complete symptom resolution, the patient presented with similar features of headache and diplopia. Similar investigative findings were seen, with cavernous sinus inflammation observed on MRI. A diagnosis of recurrent THS with isolated sixth cranial nerve palsy was established, and the patient was again prescribed prednisolone with a tapering regimen for a month. The patient was followed up for a year and reported no subsequent symptoms.

The unilateral headache and ipsilateral diplopia caused by abducens nerve palsy with cavernous sinus inflammation on MRI helped rule out the other possible causes of clinical presentation including tumors, vasculitis, basal meningitis, and sarcoid and helped establish a diagnosis of THS. The presentation fulfilled THS criteria 13.8 of the International Classification of Headache Disorders, third edition.[4]

Discussion

THS is rare, with an annual incidence of 1–2 million, with recurrence of the disease an even rarer phenomenon.[5] Our patient had relapse of symptomatology with elevated ESR and CRP levels and similar imaging presentation of cavernous sinus inflammation, helping establish the diagnosis of recurrent THS with isolated involvement of the ipsilateral sixth cranial nerve.

The existing literature reports multiple cranial nerve involvement on recurrence. Our case is the second reported case exhibiting isolated involvement of the sixth cranial nerve on recurrence.[6] Inflammation can affect single or multiple cranial nerves—oculomotor (III), trochlear (IV), trigeminal (V), and abducens (VI)—present in the cavernous sinus walls, leading to cranial nerve palsy.[3,7–10] Our case highlights the isolated involvement of a single cranial nerve, reemphasizing the importance of consideration of THS as a differential diagnosis in a similar presentation.

Steroids are considered as first-line treatment and steroid-sparing agents (cyclosporine, azathioprine, mycophenolate mofetil, methotrexate, or infliximab) as second-line or adjunct therapy for chronic pain.[6,9,10] The lack of standardized steroid dosage could be a potential reason for rare relapse of THS and necessitates further research.[11] We hope our case brings to light the rarity of single cranial nerve involvement, abducens palsy in recurrent THS to possibly ameliorate misdiagnosis of the condition. Additionally, we underscore the scarcity of research concerning the management of THS and express our hope that future investigations will delve into this area.

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