

Tolosa-Hunt syndrome

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(Key words: Tolosa Hunt syndrome)

Case report

Our patient was a 17-year-old girl who presented with a 3 week history of a sharp peri-ocular headache and drooping of the left eyelid. She also complained of a decrease in eye movement as well as dimness in vision in left eye. She indicated that she had similar episodes of headache preceded by blurring of vision in the same eye for more than two years, which responded rapidly to medications. She was otherwise healthy.

On clinical examination, her vitals were normal. She had a left sided ptosis and impairment of adduction and elevation of the left eye, consistent with oculomotor nerve palsy. The left pupil was mildly dilated but responsive to light. No facial sensory or motor loss in the distribution of the trigeminal nerve was detected. Fundoscopy was normal. Blood investigations and lumbar puncture were non-specific. The white cell count was mildly elevated and the erythrocyte sedimentation rate significantly increased.

Magnetic resonance imaging (MRI) of the brain and orbits revealed a bulky left cavernous sinus enhancing after intravenous contrast. The enhancement extended up to the left orbital apex with a downward displacement of left internal carotid artery (Figures 1 and 2). The bilateral internal auditory canals with 7th and 8th nerve complexes were unremarkable and without any evidence of cavernous sinus or superior ophthalmic vein thrombosis.

The patient was treated with oral steroids and showed significant relief of symptoms over the following 48 hours. She had complete resolution of the ptosis as well as the oculomotor nerve palsy within one month. Further follow-up showed no recurrence of symptoms.

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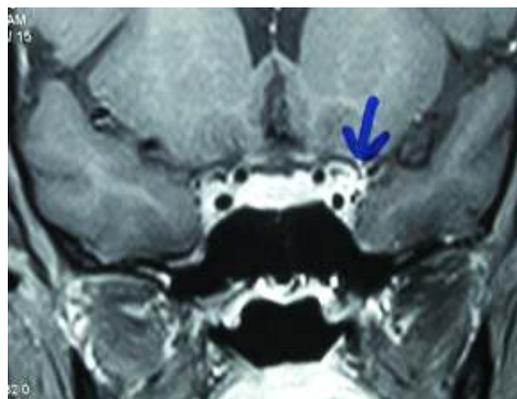


Figure 1: MRI showing bulky left cavernous sinus (indicated by arrow)

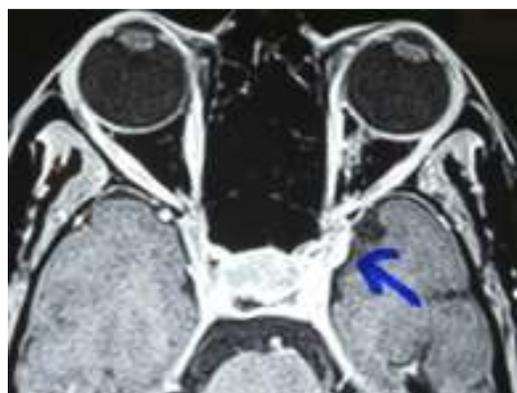


Figure 2: MRI showing bulky left cavernous sinus (indicated by arrow)

Discussion

Tolosa Hunt syndrome (THS) is a nonspecific inflammation of the septa and wall of the cavernous sinus, with lymphocyte and plasma cell infiltration, giant cell granulomas and proliferation of fibroblasts^{1,2}. The inflammation produces pressure and secondary dysfunction of the structures within the cavernous sinus, including cranial nerves III, IV, and VI, as well as the superior divisions of cranial nerve V. It is a rare syndrome with an estimated annual incidence of one case per million per year³. The diagnosis of THS requires the exclusion of other known causes of painful ophthalmoplegia, such as infection, neoplasm or vascular lesions, and is established based on the accompanying clinical symptoms and neuroradiologic findings⁴. Administration of

systemic steroids for 48 hours in a patient with THS produces a dramatic response that allows differentiation of this cause from other conditions of painful ophthalmoplegia⁵.

References

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