Tolosa-Hunt Syndrome

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Keywords

neuro-ophthalmology, clinical specialty, neuroradiology, clinical specialty, ophthalmoplegia, cranial nerve diseases, Tolosa-Hunt syndrome, cranial nerve diseases

The Neurohospitalist
2018, Vol. 8(2) 104-105
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DOI: 10.1177/1941874417714147
journals.sagepub.com/home/NHO

Figure 1. T1 axial with contrast magnetic resonance imaging (MRI) of the brain showing diffusely enhancing ill-defined enlarged left optic nerve and left medial and inferior rectus muscles, with extension into the left cavernous sinus.

A 25-year-old woman had headache and painful eye movements. Examination revealed left proptosis, ptosis, visual acuity OS (20/50), equally reactive pupils, a swollen optic disc oculus sinister-left eye (OS), and a normal optic disc oculus dexter-right eye (OD). Sensation was decreased on the left forehead. Magnetic resonance imaging revealed a diffusely enhancing retro-orbital mass extending to the superior orbital fissure and cavernous sinus (Figure 1). Steroids resulted in improvement but symptoms recurred after they were discontinued. Out of concern for malignancy a biopsy was done, showing dense fibrosis with calcified fragments (Figure 2).

Orbital pseudotumor accounts for 6% to 8% of all inflammatory lesions of the orbit with unknown etiology.^{1,2} It is often subclassified on the basis of the anatomic target areas within the orbit. Tolosa-Hunt syndrome (THS) is a granulomatous inflammatory process adjacent to the cavernous sinus or within the superior orbital fissure and/or orbital apex. The clinical diagnostic criteria of THS include an episode or episodes of unilateral orbital pain; paralysis of one or more of the third, fourth, and sixth cranial nerves; and resolution of pain within 72 hours after corticosteroid therapy, with exclusion of other causes.³

Figure 2. Biopsy showing dense fibrosis with polarized materials and calcified fragments, with no evidence of malignancy.

Magnetic resonance imaging findings of THS show inflammatory changes within anterior cavernous sinus, with or without involvement of the internal carotid artery, superior orbital fissure, orbital apex, and/or optic nerve. On the involved regions, T1 images may appear isointense with T2 hyperintensity and contrast enhancement. In a study by Schuknecht et al about 15 patients with 20 episodes of THS, the cavernous sinus is affected in the majority of patients, ipsilateral extension of the pathologic tissue into the superior orbital fissure occurred in 13 (65%) of 20 episodes, and involvement of the orbital apex occurred in 8 (40%) of 20 episodes.⁴

Although THS is often a benign condition, it may lead to optic nerve involvement and blindness. Therefore, it is important for

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neurohospitalists to recognize the symptoms and clinical features of THS and provide immediate treatment to preserve visual and ocular motility function and to prevent sequelae at a later stage.

Authors' Note

Asri Yuliati designed and drafted the article. Kumar Rajamani reviewed the manuscript and interpreted the structural MRI.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

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