Painful ophthalmoplegia: The Tolosa-Hunt syndrome

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A 49 year-old Filipino lady presented to us with 1 day history of generalized headache, vomiting, right eye pain and dizziness. There was no history of seizures, loss of consciousness, weakness or visual disturbances. She was hypertensive and on irregular medication. On examination she was conscious, alert and oriented. Pulse was 50/min, blood pressure 220/130, temperature 36.6°C. There was no focal neurological deficit, papilloedema or evidence of meningeal irritation. Hypertension was controlled with captopril and amlodipine.

On the following day, the patient continued to have right orbital pain associated with diplopia on looking to the left. She developed features of right third nerve palsy partial ptosis, with pupil sparing. Sensory impairment in the distribution of the ophthalmic division of the fifth nerve was detected on the same side. There was no proptosis or chemosis and the fundus examination was normal. There were no long tract signs. MRI findings are shown in Figures 1, 2, and 3. Figure 4 is a follow-up MRI 6 months later after treatment with corticosteroids.

Figure 1. Coronal T2W image showing mildly hypointense abnormal soft tissue in the parasellar region.

Figure 2. Axial post contrast T1W images showing enhancing soft tissue involving the parasellar region and cavernous sinuses on both sides with significantly greater involvement on the right where extension is seen to the orbital apex.

DISCUSSION

In a patient presenting with painful ophthalmoplegia, etiologies fall into three broad categories including vascular, neoplastic and inflammatory conditions. Diabetes mellitus is an important microvascular cause of painful pupil-sparing ophthalmoplegia. Tuberculosis, syphilis, mucormycosis, Herpes zoster and sinusitis are some of the infective conditions. Primary and secondary intracranial neoplasms and collagen diseases may masquerade as the Tolosa Hunt Syndrome. Intracranial aneurysms should be ruled out by MR or cerebral angiography, especially if anisocoria is detected. Essentially this syndrome remains a diagnosis of exclusion. Tolosa demonstrated granulomatous inflammation of the carotid sinus in his case at necropsy. MRI characteristically shows abnormal enhancing lesions in the cavernous sinus. Typically it is isointense with the grey matter on short TR/TE* resolution of images and iso to slightly hypointense on long TR/TE images.¹ Lateral convex bulging of the cavernous sinuses and a variable extension to the ipsilateral orbital apex, the sphenoid sinus and the floor of the middle cranial fossa and subtemporal fossa have been described. Similar signal intensity and enhancement has been described with other granulomatous lesions like sarcoidosis and tuberculosis and in lymphoma and meningiomas.²

The condition responds dramatically to cortico-

Figure 3. Coronal post contrast T1W image showing the abnormal enhancing tissue in the parasellar region causing lateral convex bulging of the cavernous sinuses.

*Time to repetition and time to echo in spin (echo sequence of MRI).

Figure 4. Post-contrast coronal T1W image showing near complete resolution of previously detected abnormality involving the parasellar region.

steroids. Pain subsides within 72 hours of initiation of corticosteroids followed by resolution of the ophthalmoplegia. Prompt response to steroids has often been used to confirm the diagnosis of Tolosa-Hunt Syndrome. However, some cases of lymphoma, sarcoidosis, Wegener's granulomatosis and intracranial aneurysms may also show partial improvement.

In 1988 Hunt and Brightman recommended that

the criteria for the diagnosis of Tolosa-Hunt Syndrome should be extended to include involvement of the 2nd and 3rd division of the 5th nerve, structures which lie outside the cavernous sinus. They also downplayed the need for histological confirmation of the diagnosis. Surgical exploration and biopsy should only be considered for very early relapses or poor response to steroids.³

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