

CASE REPORT

TOLOSA HUNT SYNDROME

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ABSTRACT: Tolosa-Hunt syndrome (THS) is a painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure. The syndrome consists of periorbital or hemicranial pain, combined with ipsilateral ocular motor nerve palsies, oculosympathetic paralysis, and sensory loss in the distribution of the ophthalmic and occasionally the maxillary division of the trigeminal nerve. Although they have relapsing and remitting course, they respond promptly to systemic corticosteroid therapy. The diagnostic eponym Tolosa-Hunt syndrome has been applied to these patients and it is this entity which forms the basis of this review.

CASE REPORT: A 42 year old man presented with painful diminution of vision since one month and diplopia in the left eye since 7 days. Tingling sensation was noted over left half of upper face.

On clinical examination -V/A was 6/12 (B/E) improving to 6/6 with pinhole, IOP and fundus was normal both eyes, extra ocular movement was completely restricted in left eye with painful movements. Left III, IV and VI Cranial nerve palsies were noted with decreased sensation over the area supplied by V Cranial Nerve.

The patient was sent for an MR examination. MR imaging was performed on a 1.5 Tesla MR scanner (GE). 10ml Gadolinium was injected in the left antecubital vein to obtain post contrast T1WI. Study revealed moderately enhancing soft tissue in the region of left cavernous sinus and orbital apex. It was isointense to muscle on T1WI and hypo intense to fat on T2WI.

MANAGEMENT: On the basis of history and imaging findings, a diagnosis of THS was made and the patient was started on corticosteroid therapy: Injection Methyl-Prednisolone (1gm.I.V for 3 days). Dramatic improvement in diplopia and pain was noted within 48 hours of starting IV steroid therapy. Oral Prednisolone 40mg daily was started after 3 days and continued for 6 weeks. At the end of therapy the patient's ophthalmoplegia also recovered.

DISCUSSION: Tolosa-Hunt syndrome (THS) is a rare disorder characterized by severe and unilateral headaches with extra ocular palsies, usually involving the third, fourth, fifth, and sixth cranial nerves, and pain around the sides and back of the eye, along with weakness and paralysis (ophthalmoplegia) of certain eye muscles¹.

The syndrome of painful ophthalmoplegia consists of periorbital or hemicranial pain, combined with ipsilateral ocular motor nerve palsies, oculosympathetic paralysis, and sensory loss in the distribution of the ophthalmic and occasionally the maxillary division of the

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trigeminal nerve. Various combinations of these cranial nerve palsies may occur, localizing the pathological process to the region of the cavernous sinus/superior orbital fissure.

The constellation of findings described may be due to four major causes: trauma, neoplasm, aneurysm, and inflammation. Comprehensive patient evaluation is essential in establishing the correct diagnosis. It may experience involvement of additional cranial nerves ipsilateral to the ophthalmoplegia, including the optic nerve, mandibular branch of trigeminal nerve, and facial nerve. Having a relapsing and remitting course, they respond promptly to systemic corticosteroid therapy. Over the past quarter century there has been no progress in understanding the pathogenesis of Tolosa-Hunt syndrome. In terms of clinical management some new information is available thanks to advances in neuroimaging techniques. Initially, radiological studies of patients with the syndrome yielded normal results, or subtle cerebral angiographic abnormalities^{2, 3, 10}. Orbital venography often disclosed abnormalities in filling of the superior ophthalmic vein or cavernous sinus, but these techniques are difficult and therefore not consistently performed. In addition, venographic abnormalities are not specific for Tolosa-Hunt syndrome, and may be found with space occupying lesions or other inflammatory processes in the orbit or parasellar region. With the advent of CT and MRI direct visualization of the cavernous sinus is possible. Using these neuroimaging modalities, cavernous sinus^{9,11} abnormalities have now been described in Tolosa-Hunt syndrome.

Clinical profile of Tolosa-Hunt syndrome

Tolosa-Hunt syndrome can affect people of virtually any age from the 1st to the 8th decades of life, with no sex predilection. Either side may be affected, with case reports of bilateral simultaneous involvement. Uniformly, patients complain of pain, which is a defining symptom. The pain lasts an average of 8 weeks if untreated. Ocular motor cranial nerve palsies may coincide with the onset of pain or follow it within a period of up to 2 weeks. It is usually described as “intense”, “severe”, “boring”, “lancinating” or “stabbing”. It is periorbital in location, frequently extending into the retro-orbital, frontal, and temporal regions. Although Tolosa-Hunt syndrome is a self limited illness, it does cause considerable morbidity. Infrequently, residual cranial nerve palsies persist. With the institution of corticosteroid therapy, the natural course is altered. Although there is no conclusive evidence to show that steroids lessen the degree or duration of the ophthalmoplegia, there is dramatic reduction of pain, often within 24 hours.

DIAGNOSTIC EVALUATION: With careful clinical examination, pain associated with typical cranial nerve palsies localises the pathological process to the regions of the cavernous sinus/superior orbital fissure. As noted above, current neuroimaging modalities allow visualization of the area of suspected pathology. Contrast enhanced MRI with multiple views, particularly coronal sections, should be the initial diagnostic study performed. Numerous reports have demonstrated an area of abnormal soft tissue in the region of the cavernous sinus in most, but not all, patients with Tolosa-Hunt syndrome^{12,13}. Typically, the abnormality is seen as an intermediate signal intensity on T1 and intermediate weighted images, consistent with an inflammatory process. In addition, there is enhancement of the abnormal area after intravenous injection of paramagnetic contrast. With corticosteroid therapy, the abnormal area decreases in volume and signal intensity in most reported cases.

High resolution CT can also demonstrate soft tissue changes in the region of the cavernous sinus/superior orbital fissure, but is less sensitive than MRI¹². This is due to lack of sensitivity to soft tissue change with superimposed beam hardening and bone streak artifacts.

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Thus, even if CT is normal, MRI must still be performed to appropriately evaluate the region of the cavernous sinus or superior orbital fissure.

Categorically, Tolosa-Hunt syndrome is a diagnosis of exclusion requiring careful patient evaluation to rule out tumour, vascular causes, or other forms of inflammation in the region of the cavernous sinus/superior orbital fissure. Neurosurgical biopsy is only rarely employed to establish the diagnosis⁴⁻⁸.

TREATMENT: Almost 40 years ago Hunt first documented the beneficial effect of corticosteroid therapy in Tolosa-Hunt syndrome.³ Unfortunately, since then there is little new information as to optimal dosage, duration of treatment, or alternative forms of therapy. It is clear that spontaneous remissions may occur, but there is no doubt that corticosteroids markedly reduce the periorbital pain. No data are available as to whether treatment hastens recovery of the associated cranial nerve palsies. Although steroids are generally tapered over weeks to months, in some cases prolonged therapy may be necessary.

DIFFERENTIAL DIAGNOSIS: Anisocoria, Arteriovenous Malformations, Benign Skull Tumors, Cavernous Sinus Syndromes, Cerebral Aneurysms, Cerebral Venous Thrombosis, Craniopharyngioma, Diabetic Neuropathy, Epidural Hematoma, Metastatic Disease to the Brain, Migraine Headache with the extra ocular muscle palsies, Neurosarcooidosis, Pituitary Tumors, Primary Malignant Skull Tumors, Tuberculous Meningitis, Varicella Zoster.

CONCLUSION: Although the pathogenetic basis of Tolosa-Hunt syndrome remains unknown, from a practical clinical standpoint it can be regarded as a distinct entity which may be simulated by various other disorders. It cannot be emphasized too strongly that patients suspected of having the syndrome require careful evaluation, appropriate treatment, and scrupulous follow up observation.

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TOLOSA HUNT SYNDROME: PICTURE GALLERY

