

CASE REPORT

TOLOSA-HUNT SYNDROME MIMICKING AS ORBITAL COMPLICATION OF SINUSITIS

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INTRODUCTION: The Tolosa – Hunt syndrome is a rare, benign condition characterized by severe unilateral headache with extraocular palsies, orbital pain caused by nonspecific granulomatous inflammation in the cavernous sinus, superior orbital fissure or the orbit.⁽¹⁻⁴⁾ The incidence of Tolosa - Hunt syndrome has been estimated as approximately one to two cases per million. The etiology of the syndrome is largely unknown and it can affect people of virtually any age, with no sex predilection. It is usually unilateral, with no predisposition for right or left side; it has been reported as bilateral in 4.1-5 % cases.^(2, 4) We report a rare case of Tolosa-Hunt syndrome which was misdiagnosed as sinusitis with orbital complication. The clinical features, diagnostic investigation and the importance of magnetic resonance imaging (MRI) studies in the differentiation of the condition are addressed.

KEYWORDS: Tolosa-Hunt Syndrome, Sinusitis, Ophthalmoplegia.

CASE REPORT: A 45-year old man was referred to our department with the suspicion of an orbital complication of sinonasal disease. The patient complained of a mild right periorbital pain, which had been diagnosed as sinusitis by a general practitioner five days earlier and treated with oral amoxicillin and clavulanic acid. On the day of admission to our department, the pain had become more intense, with the development of slight right upper eyelid oedema, right upper eyelid ptosis and horizontal diplopia of right gaze (Figure 1).



Figure 1

A complete clinical evaluation including nasal endoscopy was performed and was found to be normal. Ophthalmological and neurological examinations revealed right 6th cranial nerve palsy,

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exophthalmos, hypoesthesia in the distribution of ophthalmic and maxillary division of right 5th cranial nerve. Complete blood counts including serum electrolytes, erythrocyte sedimentation rate (ESR) and C-reactive protein were normal. Computed tomography (CT) scan revealed enhancement of right cavernous sinus. A gadolinium-enhanced MRI of the brain and skull showed enhancement of right cavernous sinus (Figure 2); it was isointense to muscle on T1W1 and hypointense to fat on T2W1. No radiological abnormalities were noted in paranasal sinuses.

Tests for angiotensin-converting enzyme, thyroid-stimulating hormone, autoimmune antibodies, antinuclear antibody (ANA), antineutrophil cytoplasmic antibodies (ANCA), extractable nuclear antigen (ENA), Antimitochondrial antibodies (AMA), rheumatoid factor and serum complement were also normal. Tuberculin skin test, chest X-ray, venereal disease research laboratory (VDRL) tests was also normal.

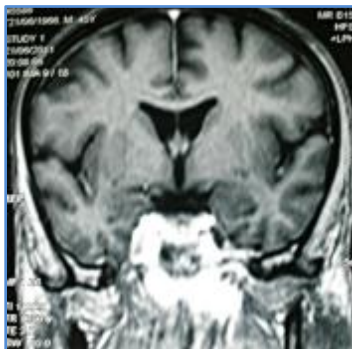


Figure 2

Corticosteroid therapy (Methylprednisolone 2 mg/kg/day) intravenous injection was commenced in two divided dosages per day and within five days all symptoms disappeared including ptosis and ocular movements (Figure 3). Subsequently after five days, patient was started on oral Prednisolone 60mg per day in three divided doses and tapered over next ten days of time. Based on all these findings, a diagnosis of Tolosa-Hunt syndrome was made. Patient was followed up for a period of six months and there was no recurrence.



Figure 3

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DISCUSSION: In 2004, the International headache society established the following diagnostic criteria for Tolosa-Hunt syndrome. Episode(s) of unilateral orbital pain for an average of 8 weeks if left untreated associated paresis of the 3rd, 4th or 6th cranial nerves which may coincide with the onset of pain followed up for a period of up to 2 weeks, pain that is relieved within 72 hours of steroid therapy, exclusion of other conditions by neuroimaging and angiography though not compulsory.⁽⁵⁾

The onset of Tolosa-Hunt syndrome is usually acute, and the patient's symptoms last for days to weeks. In 30 per cent of cases, there is sensory loss in the distribution of the ophthalmic division of the trigeminal nerve, while optic nerve dysfunction has also been reported. Rarely, the maxillary and mandibular branches of the trigeminal, facial or vestibulocochlear nerves may be affected.^(2,3) Other features include fatigue, arthralgia and feeling of protrusion of eyeball.

Due to the location and small size of the lesion in the cavernous sinus, biopsy is technically difficult and dangerous. Therefore, imaging results are highly significant in the diagnosis of the syndrome. High resolution CT scan demonstrate soft tissue changes in the region of the cavernous sinus and superior orbital fissure, but this modality is less sensitive than MRI, due to the former's lack of sensitivity in soft tissue imaging. Contrast-enhanced MRI is the examination of choice. The abnormality is seen as intermediate signal intensity on T1 and intermediate weighted images, consistent with an inflammatory process.⁽⁶⁾ Intravenous injection of paramagnetic contrast reveals enhancement of the abnormal area. It is to be noted that, a small number of patients show normal MRI findings.^(2,3)

The MRI findings before and after systemic corticosteroid therapy are important in the definitive diagnosis of Tolosa-Hunt syndrome.^(2,7) MRI with 3-dimensional constructive interference in steady state (3D CISS) provides an enhanced picture within the cavernous sinus.⁽⁸⁾ This type of imaging may assist with the future diagnosis, but it is not yet used routinely. However, diagnostic MRI findings do not as yet exist, making Tolosa-Hunt syndrome a diagnosis of exclusion. Therefore careful patient evaluation is required in order to rule out tumors, vascular causes, infection and other causes of painful ophthalmoplegia (Table 1).^(2,3)

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TABLE 1

Caption: CAUSES OF PAINFUL OPHTHALMOPLEGIA (2)

1. Trauma
2. Tumors (Pituitary adenoma, Craniopharyngioma)
3. Vascular
 - Intracavernous carotid artery aneurysm
 - Posterior fossa aneurysm
 - Carotid-cavernous fistula
 - Carotid-cavernous thrombosis
4. Infections (TB meningitis, Lyme disease, Whipple's disease, Varicella zoster)
5. Tolosa-Hunt syndrome
6. Neurosarcoidosis
7. Eosinophilic granuloma
8. Orbital diseases
9. Diabetic ophthalmoplegia
10. Basal meningitis
11. Vasculitis
12. Ophthalmoplegic migraine

Although ESR is expected to be elevated in chronic granulomatous diseases, it is not so in the present case, probably due to the short duration of the disease.

Tolosa-Hunt syndrome is essentially a benign disorder which responds rapidly to the administration of corticosteroids. However, some cases do not respond to corticosteroids and there is also evidence of spontaneous remissions. Recurrences are common, which have been observed in 50% of reported patients, within months or years of the initial attack. Patients who have experienced spontaneous remission appear to have as much risk of recurrence as those treated with medication. Although Tolosa-Hunt syndrome is a self-limiting illness, still it is known to cause considerable morbidity. Rarely residual cranial nerve palsies may persist. Orbital, periorbital pain and paresis resolve within 72 hours when treated adequately with corticosteroids.⁽²⁾ The disappearance of symptoms following systemic corticosteroid treatment may precede normalization of neuroradiological studies by weeks or even several months.⁽²⁾

On the other hand the clinical improvement after steroid therapy is not absolute proof of diagnosis, as lymphoma, meningioma and giant cell tumours also respond symptomatically to steroids, albeit with some delay; however, signs do not resolve.⁽⁹⁾ For refractory cases, Azathioprine, Methotrexate or radiation therapy has been employed. Therefore we believe that follow-up MRI studies in these patients are vital, in order to monitor evolution of the lesion and also to avoid an incorrect diagnosis of Tolosa-Hunt syndrome in the presence of other lesions.

To conclude, although Tolosa-Hunt syndrome is rare, a high degree of suspicion is needed in patients with suspected sinusitis with oculomotor muscle palsy not responding to the initial line of management for sinusitis. In such cases MRI should be the investigation of choice both for diagnosis and follow up.

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