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A Patient Presenting with Ptosis, Ophthalmoplegia, and Decreased Periorbital Sensations and Facial Droop in Tolosa-Hunt Syndrome

Ali Hasnie MD*¹, Usman Hasnie MS-IV², Mudassar Zia, MD^{2,3}

¹Mayo Clinic-Rochester, MN; ²University of Missouri-Kansas City School of Medicine; ³Truman Medical Center

Abstract

Introduction: Tolosa-Hunt Syndrome is a diagnosis of exclusion because a single characteristic pathognomonic finding for this condition does not exist. Although painful ophthalmoparesis or ophthalmoplegia is the hallmark of this syndrome, it is an infrequently reported entity. Even more infrequent is the occurrence and reporting of Tolosa-Hunt Syndrome with involvement of a cranial nerve seven (facial nerve). In fact, in our literature review we were able to find only one such documented case.

Case presentation: We present a case of Tolosa-Hunt Syndrome in a twenty-nine-year-old Hispanic male patient admitted to our hospital for left sided ptosis, ophthalmoplegia, facial droop, decreased periorbital sensations associated with severe ipsilateral stabbing peri-orbital pain and headache. He was managed on high dose steroids and improvement was noticed within 24 hours.

Conclusion Tolosa-Hunt Syndrome is an extremely painful condition that responds well to high dose steroids. However, due to the fact that it is so uncommon and a diagnosis of exclusion, it is often overlooked. We hope this report will assist both the general internist and the family practitioner to consider this diagnosis among their differential when a patient presents with these features.

Introduction

Tolosa Hunt syndrome, first described by Tolosa in 1954, is a rare condition occurring in approximately one person per million people per year (1). Pathologically, it is defined by idiopathic granulomatous inflammation of the cavernous sinus or supra-orbital fissure, commonly unilateral but sometimes bilateral. Clinically it presents with acute onset severe

periorbital headache with extraocular palsies usually involving the third, fourth, fifth and sixth cranial nerves. Diagnosis is challenging due to the fact it is a diagnosis of exclusion, but once established it responds well to high dose steroid therapy. Relapses are common.

Case Presentation

A twenty-nine year old Hispanic male presented with left-sided ptosis, ophthalmoplegia, facial droop and decreased periorbital sensations, associated with ipsilateral severe stabbing periorbital pain and headache. About one month prior the patient noticed a spot on the medial conjunctiva of his left eye, accompanied by redness and irritation and experienced binocular horizontal diplopia. He developed severe intermittent left sided periorbital pain not relieved by high doses of Ibuprofen (twelve pills per day) that worsened when in a prolonged supine position. Two weeks later he experienced the onset of ptosis, paraesthesia of the periorbital region and paresis of the left side of his face accompanied by frontal headaches causing him to wake up several times during the night. The patient denied having any dysarthria, dysphagia, blurriness or loss of vision. He had no history of orbital trauma. The patient did experience left-sided facial weakness a year prior and was treated with an injection of morphine at a local hospital. The episode resolved within twenty four hours and he was not symptomatic during the interim period.

On physical examination the patient was alert and oriented. Left ptosis and ophthalmoplegia with severe limitation of gaze in all directions was suggestive of a deficit involving cranial nerves III, IV and VI. The patient had left lagophthalmos and superficial keratopathy. Both pupils were 4-5 mm, however the left pupil was sluggishly reactive. Visual fields were full to confrontation, visual acuity was 20/20 in the right eye and 20/40 in the left eye. A dilated fundoscopic exam of the left eye was reported normal by a neuro-ophthalmologist with no evidence of hypertensive retinopathy or diabetic retinopathy. (A neuro-ophthalmologist consultation was obtained in favour of a general ophthalmology consultation.) Sensation was intact to light touch, pin-prick and temperature on the right side of the patients face, but was notable for diminished sensation on entire left side of patient's face. Corneal reflex was present (i.e. ipsilateral and consensual). In addition, the patient had cranial nerve VII deficit with paralysis of the entire left side of the face as well as facial droop. The patient was unable to close the left eye when instructed to nor raise his left eyebrow but was able to raise his right eyebrow. He was unable to smile with the left side of his face. This paralysis demonstrated by the patient is consistent with a lower motor neuron injury of Cranial Nerve VII, and consistent with the House-Brackmann Grade 5 facial paralysis. Lab findings were unremarkable, including infectious workup with a lumbar puncture (LP). CT of the head without contrast showed no acute intracranial abnormality. MRI of brain and orbits revealed asymmetric abnormal enhancement along the left cavernous sinus with enhancing fullness on the left extending from Meckel's cave to the area of the foramen rotundum and mild thickening and enhancement of the adjacent duramater extending into the greater wing of the left sphenoid bone. There was also subtle enhancement of the left optic nerve near the orbital apex and asymmetric enhancement along the left orbital apex. Constellation of findings was non-specific but suggestive of idiopathic inflammatory pseudotumor. The patient was treated with 250 mg

prednisolone IV for five days resulting in dramatic improvement in his symptoms and then continued on 85 mg oral prednisone taper therapy over the course of two weeks. He was advised to follow up in the eye clinic in two weeks. Follow up revealed significant improvement of cranial nerves III, IV, and VI. Cranial nerve VII improvement was less significant than the other cranial nerves. The patient was to follow up 3-4 weeks following this appointment but was lost to follow up, presumably due to continued improvement.

Of note, the major differential diagnosis in this patient for our team was between Tolosa-Hunt syndrome and Cavernous sinus thrombosis. The key in our management was to effectively rule out cavernous sinus thrombosis as this condition is a life threatening condition which could have been propagated with the initiation of steroids. The patient's vitals demonstrated that there was no point in time at which he was febrile. On exam, there was no proptosis, or chemosis noted at any time during the clinical course. Radiologic imaging (including CT and MRI) was found to be inconsistent with cavernous sinus thrombosis as the lateral wall of the cavernous sinus did not demonstrate convex margins and there was no dilation of the ipsilateral superior ophthalmic vein. Laboratory work did not demonstrate a notable increase in white blood cell count, and CSF analysis was negative for infectious etiology including *Borrelia burgdorferi*. And finally, treatment with high dose steroids resulted in dramatic improvement in the patient, which ruled out cavernous sinus thrombosis.

MRI Images:

Figure 1

Figure 2

tolosa-hunt-figures-1-2

The image on the left (figure 1) demonstrates the area of asymmetric enhancement of the cavernous sinus with the red arrow indicating the area of abnormal enhancement initially concerning for cavernous sinus thrombosis on an axial MRI.

The image on the right (figure 2) is a coronal MRI demonstrating the area of abnormal enhancement with the red arrow indicating the area of enhancement.

Clinical Images:

tolosa-hunt-figure-3

Figure 3. This image demonstrates the patients attempted gaze to the right. Notice the absence of movement of the left eye.

tolosa-hunt-figure-4

Figure 4. The image above demonstrates attempted gaze to the left. Notice the absence of movement of the left eye.

tolosa-hunt-figure-6

Figure 5. The patient is attempting to raise his eyebrows. Notice the lack of movement of the left eye. The patient also had asymmetrical facial expression grossly. This is consistent with a Grade 5 House-Brackmann classification of facial nerve function.

Discussion

Although Tolosa-Hunt syndrome is infrequent, it may present at any age with an estimated incidence of one per million (1, 2, 3), and the diagnosis should be entertained in patients presenting with painful ophthalmoparesis or ophthalmoplegia accompanied by periorbital pain and a varying constellation of ipsilateral oculomotor cranial nerve palsies. According to Hunt's diagnostic criteria (2), the clinical course of Tolosa-Hunt syndrome is unpredictable. Onset is usually acute and symptoms may persist for days or weeks before resolving spontaneously, sometimes with residual neurologic deficit. The condition however follows a relapsing and remitting course; symptoms may recur within months or years of the initial episode (4, 5) with an ipsilateral, contralateral or even bilateral presentation (6). Although the etiology is obscure, the pathological process is a non-specific inflammation. Proliferation of fibroblasts and infiltration of the septa and wall of the cavernous sinus with infiltration of the septa and wall of the cavernous sinus in the absence of necrosis exert pressure upon the penetrating nerves resulting in the varying degrees of neurological involvement (7). Systemic symptoms have not been reported as a part of the entity. Due to the fact that THS is a diagnosis of exclusion, other causes of painful ophthalmoplegia must be carefully considered. Trauma, vascular, neoplastic and infective causes as well as orbital disease, diabetic ophthalmoplegia, and posterior fossa aneurysms must be ruled out. High resolution CT and MRI are the neuroimaging modalities that allow for visualization of the pathology and evaluation of the cavernous sinus region; even though lack of specificity of the findings is a limitation. Spontaneous remission of the condition is the usual course but steroids have been well documented to result in marked reduction of the periorbital pain, usually within 24-48 hours. In fact, Hunt first documented the beneficial effect of corticosteroid therapy in THS (2).

Summary

Tolosa-Hunt Syndrome is an extremely painful condition that responds well to high dose steroids. However, due to the fact that it is uncommon, a diagnosis of exclusion and sparsely documented, the diagnosis may be overlooked.

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