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Case Report

Tolosa Hunt Syndrome in A 45 Year Old Male Presenting as Headache

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Abstract

Background: Tolosa Hunt Syndrome is a rare condition caused by an idiopathic inflammation of the cavernous sinus responsive to steroid treatment.

Case Report: A case of a 45 year old previously healthy man whose chief complaint was headache and painful opthalmoplegia. Neurologic examination on admission revealed left ptosis, miotic but reactive pupil, and left ophthalmoplegia. Ophthalmic division of cranial Nerve V revealed an 80% sensory loss. Localization was on the left cavernous sinus. MRI revealed subtle asymmetrical enhancement of the left cavernous signs which is suggestive of inflammation. He was given an oral course of steroids with marked resolution of symptoms after two months with no recurrence after one year on follow up via telemedicine.

Conclusion: The diagnosis of Tolosa hunt was based on clinical grounds. Ancillary testing (other than cranial imaging) returned normal excluding other considerations. This was coupled with exquisite response to steroids. We agree that tissue biopsy remains the standard to make the diagnosis with certainty although not practical for some patients. Surgical biopsy is reserved for symptoms that do not respond promptly to steroids and if they recur after the initial honeymoon period with steroids has lapsed.

Keywords: Tolosa Hunt Syndrome; Etiology; Surgical Biopsy

Introduction

Tolosa Hunt (TH) syndrome is defined by an idiopathic inflammation of the septa and wall of the cavernous sinus. It is a rare syndrome with an estimated case of one case per million per year and the etiology of which remains unknown up to today [1]. First defined in the year 1945; this condition is renowned for its favorable response to corticosteroid treatment [2]. The syndrome begins as a generally mild to moderately worrisome headache progressing with or concurrent to painful ophthalmoplegia. If left untreated it is, self-limiting after 8 weeks. The International Headache Society defines the diagnostic criteria for TH syndrome [3]. There is no single test that will identify TH syndrome and it remains a diagnosis of exclusion. We report a 45-year-old man who presented with the classic description of this disease, workup, and treatment.

Case

A 45-year-old previously healthy man presented at the outpatient department for a chief complaint of insidious onset of diplopia. Preceding three weeks prior, he experienced throbbing and occasionally gnawing moderatel intense headache localized at the back of his left eye with radiation to the temporal area. A tingling sensation was noted on the left temporal area and left ophthalmoplegia followed suit. No associated symptoms such as

blurred vision, vomiting, dizziness, loss of consciousness or limb weakness. He has no history of sexually transmitted disease. Family history was non contributary and he was not on any maintenance medications.

Neurologic examination on admission revealed left ptosis, miotic but reactive pupil, and left ophthalmoplegia. The right eye had normal ocular movements, visual fields and acuity. Fundoscopy was normal for both eyes. The ophthalmic division of cranial Nerve V revealed an 80% sensory loss. The rest of the neurologic and systemic physical examination was unrevealing,

A hematologic exam revealed mild leukocytosis (WBC of 11000/ml) and a normal hemoglobin and platelet count. The coagulation exam was also within limits. Liver and renal function tests did not reveal any metabolic perturbations. ESR and CRP did not exceed the reference range stated. Serum ANA, anti SM, anti DSDNA, CEA, AFP also were below reference range values. A CSF analysis was done showing normal opening pressure, zero RBC, slight elevation of WBC (5) with 100% lymphocytic predominance, no organisms were seen on gram stain and grown on culture. CSF was negative for oligoclonal bands and VDRL was non-reactive.

Figure 1: Left ophthalmoplegia.

An MRI was done to this patient showing that a subtle asymmetry of the cavernous sinus was appreciated, left more prominent than the right which was suggestive of an inflammatory process. MRA of the brain demonstrated normal appearance of the distal vertebral, basilar, and intracranial carotid arteries. The anterior, middle, and posterior cerebral arteries are normal bilaterally. No obvious areas of vascular narrowing or dilatation were identified.

Since the patient's neurologic status remained stable while we performed these diagnostic exams concurrently, definitive treatment was delayed until a proper investigation was completed. After reaching a consensus of TH syndrome – with agreement from Neuro-Ophthalmology service; the patient was started on oral prednisone (1 mg/kg/day) in three divided doses for 2 weeks. We deferred giving methylpredinosolone pulse for this patient and was the preference for both neurology and ophthalmology service. Appropriate gastroprotective measures were given and patient was advised of steroid related adverse effects.

Follow up after two weeks showed marked improvement of the left ophthalmoplegia. Patient now demonstrated abduction, adduction, intorsion and extortion of the left eye. Ptosis was diminishing. He was given a steroid tapering schedule for three months. He returned for his second follow up on the second month post discharge and was advised to repeat the cranial MRI on the second quarter of the year. Unfortunately, due to the covid pandemic, stay at home orders were in place and a repeat cranial imaging was deferred by the patient at this time.

He later reappeared in our telemedicine consult service after one year for follow up. Limited neurologic examination delivered via telemedicine showed full extraocular movements of both eyes with no recurrence of the painful ophthalmoplegia. He was advised to continue to monitor for any neurologic symptoms that may reappear in the future.

Discussion

This case report demonstrates a neurologic lesion with dramatic response to steroids. Based on histopathologic studies of samples taken from patients with similar presentation - the inflamed wall and septa of the sinus demonstrates fibroblast proliferation, lymphocytic and plasma cell infiltration, and giant cell granuloma formation [4]. Proliferation of cells causes mass effect, pressure impingement of cranial nerves III, IV, VI and superior division of Cranial nerve V.

The clinical profile and temporality is not inherently unique for Tolosa Hunt Syndrome since other conditions mimic it. More than 75% of patients presenting with painful opthalmoplegia do not have Tolosa Hunt syndrome [6]. Our differential diagnosis in the case were structural compressive lesions that included the following: primary tumor, metastatic tumor, lymphoma, meningioma, aneurysm, cartotid cavernous fistula, carotid dissection, cavernous thrombosis, vasculitis, sarcoidosis and neurosyphilis and other indolent infections. Tumors are one of our main concerns since

some of these entities respond to steroids (e.g. lymphoma) and improvements in the patient's symptoms may mask the diagnosis. Completely missing out a diagnosis of intracranial aneurysm or fistula can also be fatal in the long run. To add from these structural compressive lesions are those that may be nonstructural in nature such as arteritis, ophthalmoplegic migraine, diabetic nerve palsy and demyelination.

An MRI done to this case only revealed subtle asymmetry on the left cavernous sinus – (hypointense relative to fat and isointense with muscle on T1 weighted images; isointense with fat on T2 weighted scan). No mass lesions of any kind visualized and angiography did not reveal pathologies in the intracranial internal carotid. A serial MRI may have been done in order to totally rule out the possibility of lesions that would be too small to be seen in the initial MRI. Nevertheless, in the absence of radiographic evidence of a structural cause; idiopathic inflammation seems to be more likely. Negative tumor markers supplanted evidence that malignancy was less likely. Serologic test for ANA, antiSM, antiDSDNA and ANCA were done to detect for concomitant lupus, connective tissue disease and vasculitis. CSF analysis was done to exclude the possibility of infection (bacterial and fungal meningitis, neurosyphilis) and to evaluate the presence of oligoclonal bands. These ancillary diagnostic tests were unremarkable and thus noncontributory. A surgical biopsy of the cavernous tissue would have been the most ideal – but not very practical and we opted to do a trial of steroids. Reserving this invasive procedure if the therapeutic trial fails.

We started with prednisone (1 mg/kg/day in three divided doses) for two weeks with decision to taper based on response. After 10 days, there was improvement of the ophthalmoparesis with disappearance of temporal headache and numbness. He was discharged thereafter with instructions for close follow up. After two weeks there was complete regression of signs and symptoms. The steroids were tapered down and discontinued after a total duration of three months. The plans were for a repeat MRI. Similar case reports noted normalization of MRI findings concurrent with the disappearance of signs and symptoms [7]. There were also cases of symptoms relapsing during steroid taper thus the importance of follow up and serial radiologic studies. Unfortunately for this case, he was lost to follow up due to the COVID pandemic and there was limited access to health care resources at that time. He eventually reappeared after one year in our telemedicine service with no recurrence of symptoms.

Conclusion

In our patient, the diagnosis of Tolosa hunt was based on clinical grounds. Ancillary testing (other than cranial imaging) returned normal excluding other considerations. This was coupled with exquisite response to steroids. Even with stringent adherence to clinical criteria and diagnostic evaluation, an underlying neoplasm may be missed. In our opinion doing a surgical biopsy may be too invasive for these patients. We reserve it only if symptoms do not respond promptly to steroids or symptoms reappear during steroid taper. For our patient, our conclusion is that of Tolosa Hunt Syndrome.

Bibliography

1. Iaconetta G., *et al*. "Tolosa-Hunt syndrome extending in the cerebello-pontine angle". *Cephalalgia* 25 (2005): 746.

2. Amrutkar C and Burton EV. "Tolosa-Hunt Syndrome". 2020 Aug 10. In: StatPearls. Treasure Island (FL): StatPearls Publishing (2021).

3. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia 38 (2018): 1.

4. Kline LB and Hoyt WF. "The Tolosa-Hunt syndrome". *Journal of Neurology, Neurosurgery and Psychiatry* 71 (2001): 577-582.

5. Zhang X., *et al*. "Validation of ICHD-3 beta diagnostic criteria for 13.7 Tolosa-Hunt syndrome: Analysis of 77 cases of painful ophthalmoplegia". *Cephalalgia* 34 (2014): 624.

6. Cakirer S. "MRI findings in the patients with the presumptive clinical diagnosis of Tolosa-Hunt syndrome". *European Radiology* 13 (2003): 17.

7. Zhang X., *et al*. "Factors that influence Tolosa-Hunt syndrome and the short-term response to steroid pulse treatment". *Journal of the Neurological Sciences* 341 (2014): 13.