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Tolosa-Hunt Syndrome
A Case Report and Review of the Literature

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Abstract

The Tolosa-Hunt Syndrome (THS) refers to a painful recurrent ophthalmoplegia with or without associated sensory changes. Attacks may last days to weeks and are characterized by remission and periodic relapses. THS closely resembles pathologic process(es) around the cavernous sinus or the superior orbital fissure and must be differentiated from them. The case of a young female with THS is described below followed by a review of the literature.

Key Word: *Tolosa-Hunt Syndrome*

Case Report:

G.P., a 26 year old Spanish speaking Puerto Rican female was seen in our emergency room with a 24 hour history of agonizing right orbital and right temporal pain associated with photophobia, epiphora and blurred vision of the right eye. The pain was "boring" in character and constant. The patient also complained of severe nausea and frequent vomiting. The patient was seen in a medical clinic where she was diagnosed to have migraine and was started on propanolol 40 mg tid, and minor tranquilizers, with no apprent relief. Review of her medical history was unremarkable except for recurrent attacks of right temporal headaches for four months.

Physical examination at the time of her presentation revealed an average built female who was in obvious pain with moderate tenderness over the right orbit and eyeball. She had ptosis of the right eye and oculomotor nerve paralysis with normal pupillary reaction. She also had paresthesia and hyperesthesia in the distribution of the right trigeminal nerve. Corneal reflex, ophthalmoscopic examination of the eye and ENT evaluation were all normal. The rest of the systemic examination was also normal.

The patient was initially evaluated for inflammatory and/or vascular pathology in and around the orbit. Computer tomography of the brain and overlapping cuts of the orbit with and without contrast, CSF examination and cerebral angiography were all normal. Conventional treatment of the pain afforded no relief. Prednisone was started at 60 mg qd with remarkable improvement of her orbital pain in 24 hours. During the ensuing 48 hours there was significant improvement in the patient's third nerve palsy. She was discharged from the hospital five days after the initiation of prednisone treatment with almost complete relief of her symptoms and the ophthalmoplegia. Prednisone was stopped two weeks after discharge. The patient remained asymptomatic for over ten months when she again had a similar episode, however, with less severe pain and ptosis. The patient was treated on an outpatient basis with 10 mg of prednisone qd. She again demonstrated a complete recovery in three days and prednisone was discontinued in two weeks. Since this episode the patient has remained symptom free for over a year.

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Table I
Pathological Processes Presembling THS (Tolosa-Hunt Syndrome)

Site	Pathological Process
Orbit	Contiguous sinusitis
	Mucormycosis and other fungus infections
	Metastatic tumor
	Lymphoma
Superior Orbital Fissure	Nonspecific granulomatous inflammation (Tolosa-Hunt Syndrome)
	Metastatic tumor
	Nasopharyngeal carcinoma
	Lymphoma
	Herpes Zoster
	Carotid cavernous fistula
Parasellar Area	Cavernous sinus thrombosis
	Pituitary adenoma
	Intracavernous aneurysm
	Metastatic tumor
	Nasopharyngeal sinus muocele
	Meningioma, chordoma
	Petrositis (Gravenigo's Syndrome)
Posterior Fossa	Posterior communication artery aneurysm
	Basilar artery aneurysm
Miscellaneous	Diabetic ophthalmoplegia
	Migrainous ophthalmoplegia
	Cranial arteritis

Discussion and review of literature

There are a large variety of pathological processes that may present with a clinical picture resembling THS. (Table I) It is mandatory that all relevant evaluations be undertaken to exclude these. In his original case described 35 years ago, Dr. Tolosa described a 47 year old man with left orbital pain and ophthalmoplegia with a segmental narrowing of the carotid siphon.[1] This patient underwent a left transfrontal intradural exploration and subsequently expired on the third postoperative day. The only post-mortem finding was in the intracavernous portion of the left carotid artery, which was wrapped in granulomatous tissue without intradural obstruction. Hunt et al in 1961 described six cases and proposed criteria for what is now known as the THS.[2] These criteria include: i) retro-orbital or orbital pain preceding ophthalmoplegia by days or weeks; ii) the pain typically being boring or gnawing in nature; iii) involvement of cranial nerves passing through cavernous sinus (3rd, 4th, 5th and/or 6th); iv) symptoms lasting for days or weeks; v) negative etiologic evaluations. These characteristics have been noted by many other authors and our case also fits this description.

There have been many cases of painful ophthalmoplegia with trigeminal nerve involvement described n the literature.[3-5] In one case the sensory disturbance was confined to the maxillary division,[6] but most often it is the ophthalmic branch that is involved. The region where the carotid artery enters the cranial cavity, by penetrating the outer wall of the cavernous sinus, is the site of a non-specific low grade inflammation, which has been confirmed by various authors.[1-4] However, there is no evidence that the primary process is an arteritis or manifestation of a systemic disease, although a few cases have been associated with some elevation of the sedimentation rate and anti-nuclear antibodies.[6,7]

Remissions and relapses of painful ophthalmoplegia would be highly suggestive of THS. This diagnosis, however, should be one of exclusion and every attempt should be made to exclude a more treatable and potentially fatal cause. If the condition has been observed bilaterally the diagnosis of THS can be made more confidently.[6] CT scanning has not been of much help although arteriography has frequently shown narrowing of the internal carotid artery in the cavernous sinus. Some authors have reported that orbital venography has demonstrated

impaired flow or occlusion of the superficial ophthalmic vein.[3,9,10] This could be dangerous as many conditions, some potentially treatable, like parasellar chordoma,[10] malignant lymphoma[11] and actinomycosis,[6] have shown partial or complete initial response to steriods. It would be erroneous to rely too heavily on the response to corticosteroid therapy as a diagnostic criterion.

It is appropriate to emphasize the close resemblance of THS to many treatable parasellar pathologies. An accurate diagnosis is contingent upon relevant evaluations, including CT scan and cerebral radio-contrast studies, rather than dwelling on the response to corticosteriods as a therapeutic test.

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References:

1. Tolosa, Edwardo: Periateritic lesions of the carotid siphon with the clinical features of a carotid infrasphenoidal aneurysm. J Neurosurg and Psych 1954; 17:300-302.
2. Hunt W E, Meagher J N, et al: Painful ophthalmoplegia. Neurol 1961; 11:56-62.
3. Smith J L, Taxdal D S R: Painful ophthalmoplegia. The Tolosa-Hunt Syndrome. Am J Ophthal 1966; 61:1466-1472.
4. Cheah J S, Ransome G A: Colliers syndrome. Med J Austral 1970; 1:277-278.
5. Sandheimer F K, Knapp M D: Angiographic findings in the Tolosa-Hunt Syndrome: Painful Ophthalmoplegia Radiology 1973; 106:105-112.
6. Dornan M L, Espir E A, et al: Remittent painful ophthalmoplegia: The Tolosa-Hunt Syndrome. J Neurol Neurosurg and Psych 1979; 42:270-275.
7. Mathew N T, Chandy J: Painful ophthalmoplegia. J Neurol Sciences 1970; 11:243-256.
8. Milstein B A, Morretin L B: Report of a case of sphenoid fissure syndrome studied by orbital venography. Am J Ophthal 1971; 72:600-603.
9. Mathew N T: Painful ophthalmoplegia. In Tropical Neurology) London: Oxford University Press, 1972.
10. Thomas L E, Yoss R E: The parasellar syndrome: Problems in determining etiology. Mayo Clin Proc 1970; 45:617-623.
11. Fowler T J, Earl E T, et al: Tolosa-Hunt Syndrome: The dangers of an eponym. Brit J Ophthal 1975; 59:149-154.