Case 16092

Eurorad ••

Tolosa-Hunt Syndrome

Published on 15.11.2018

DOI: 10.1594/EURORAD/CASE.16092 ISSN: 1563-4086 Section: Neuroradiology Area of Interest: Neuroradiology brain Procedure: Imaging sequences Procedure: Contrast agent-intravenous Procedure: Diagnostic procedure Imaging Technique: MR Special Focus: Pathology Motility Case Type: Clinical Cases Authors: María José Zubeldia, Bárbara Zannini, Leonel Migliacci, Maximiliano Matteoda, María Maira Fernández. Patient: 32 years, male

Clinical History:

32-year-old male patient suffering retro-orbital and right hemicranial pain without improvement with non-steroidal analgesics. He subsequently developed diplopia, progressive paresis of the right eye to outward movement and hypoesthesia in homolateral fronto-orbital area. The eye exam and laboratory tests were normal. All symptoms significantly remitted with steroid. (Figure 1)

Imaging Findings:

The brain MRI showed a moderate convex thickening of the dural edge in the right cavernous sinus extending to the superior orbital fissure, isointense in T1 and with homogeneous enhancement with gadolinium. The injury was in close contact with the intracavernous portion of the internal carotid artery, narrowing it slightly. (Figure 2) In the post-therapy control MRI, a clear reduction of the described findings was observed.(Figure 3) **Discussion:**

The Tolosa-Hunt Syndrome (THS) is a painful ophthalmoplegia caused by an idiopathic granulomatous inflammation of the cavernous sinus and/or superior orbital fissure.[1]

This syndrome affects both sexes, having the highest incidence towards the fourth decade of life.[2] Although the etiology is unknown, there are reports describing its possible autoimmune origin due to the positivity for ANCA, antilupic and antiperoxidase antibodies in some patients with THS.[1]

The typical symptoms are retro or periorbital unilateral pain with oculomotor progressive paresis due to pair III, VI and IV involvement. The pair V ophthalmic branch may be affected, producing periorbital paresthesias. Infrequently, a Horner's Syndrome can occur due to the involvement of periarterial sympathetic fibers.[3]

The implication of the image methods lies in excluding other etiologies of the cavernous sinus syndrome, showing the typical THS findings and monitoring the post-therapeutic response.[4] The method of choice is the MRI (92% sensibility).[2] There is disparity in the literature about the utility of MRI diagnosis criteria.[5] The MRI findings consist of a convex thickening of the cavernous sinus that may extend to the superior orbital fissure and orbit, with a variable intensity in T1 and T2 and homogenous enhancement.[5, 6]

Angiography identifies narrowing or segmental irregularities in the internal carotid artery, superior ophthalmic vein occlusion or nonvisualization of the affected cavernous sinus. However, a normal angiography does not exclude the diagnosis.[7] These alterations usually improve after corticoid therapy.

The most important differential diagnoses are sarcoidosis, lymphoma, meningioma and metastases. Sarcoidosis and lymphoma are illnesses with systemic involvement. Both respond to steroid therapy. Sarcoidosis is usually associated with multiple hypointense T2 focuses in the brain parenchyma and meninges and may cause the thickening of the cranial nerves. Unlike TSH, lymphoma involves the arteries without narrowing them. [8] The meningioma has homogenous enhancement and arterial narrowing too, but it does not respond to corticoid therapy and it may have calcifications, reactive hyperostosis and dural tail.

Metastases have a clinical history of malignancy, they are usually multiple and does not improve with steroid. A controversial differential diagnosis is the orbital pseudotumor because they share clinical-therapeutical features, the difference is that the pseudotumor would affect the orbit and the extraocular muscles.[3, 7] Differentiating TSH from other diseases can be done considering clinical presentation, neuroimaging studies (especially MRI with contrast) and the response to steroids. However, clinical and radiological improvement with steroid therapy is not specific for TSH.

Written informed patient consent for publication has been obtained. **Differential Diagnosis List:** Tolosa-Hunt Syndrome, Sarcoidosis., Lymphoma., Meningioma., Metastases.

Final Diagnosis: Tolosa-Hunt Syndrome

References:

Martínez DF, Casasco JP, Pendre N, De Bonis C, Berner SI (2010) Síndrome de Tolosa-Hunt. Revista argentina de neurocirugía 24(3), 111-115

Díaz C, Aedo I, González-Hernández J (2009) Tolosa Hunt Syndrome: A review from a clinical case. Memoriza.com 5: 1-7

Torales M, Olivera MN, Olazarri A, et al. (2012) Oftalmoplejia dolorosa, un desafío diagnóstico: a propósito de un caso clínico de Tolosa-Hunt. Med Interna 34(2):60-3

Guedes BVS, da Rocha AJ, Zuppani HB, da Silva CJ, Sanvito WL (2010) A case review of the MRI features in alternating Tolosa-Hunt syndrome. Cephalalgia 30(9): 1133–1136 (PMID: 20713564)

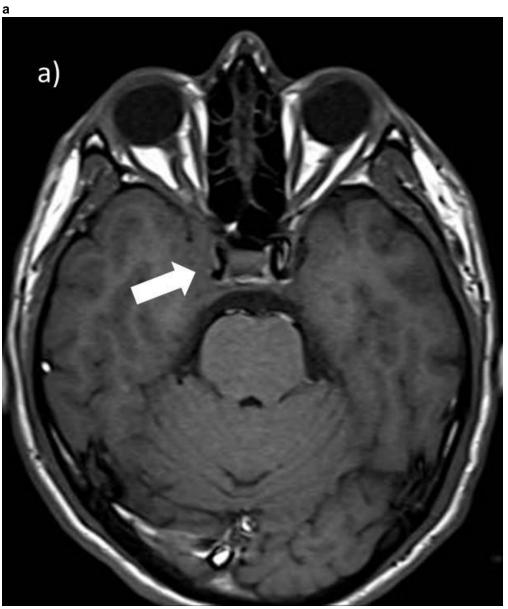
Aguirre V. D, Zúñiga G, Barrera LI (2014) Tolosa-Hunt Syndrome: Case Report And Literature Review. Acta Neurol Colomb 30(4):346-352

Sánchez Vallejo R, Lopez-Rueda A, Olarte A. M., & San Roman L, (2014) MRI findings in Tolosa-Hunt syndrome (THS). BMJ Case Reports (PMID: <u>25368129</u>)

Yousern DM, Atla SW, Grossman RI, Sergott RC, Savino PJ, Bosley TM. (1989) MR imagin of Tolosa-Hunt Syndrome. American Journal of Neuroradiology 10:1181-1184

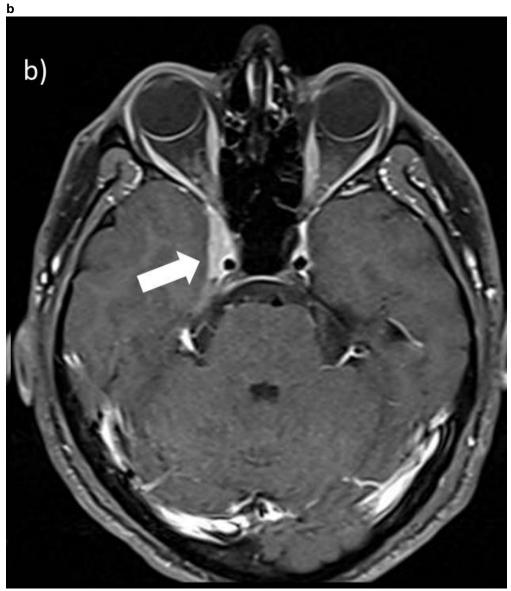
Abdel Razek AAK., Castillo M. (2009) Imaging Lesions of the Cavernous Sinus. American Journal of Neuroradiology 30 (3) 444-452

Figure 1

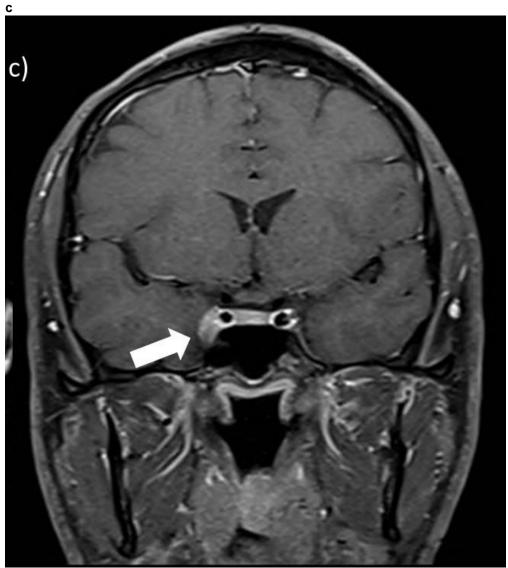


Description: a)Axial T1 SE

Dural thickening of the lateral border of the right cavernous sinus with extension to the superior orbital fissure (white arrow) **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina

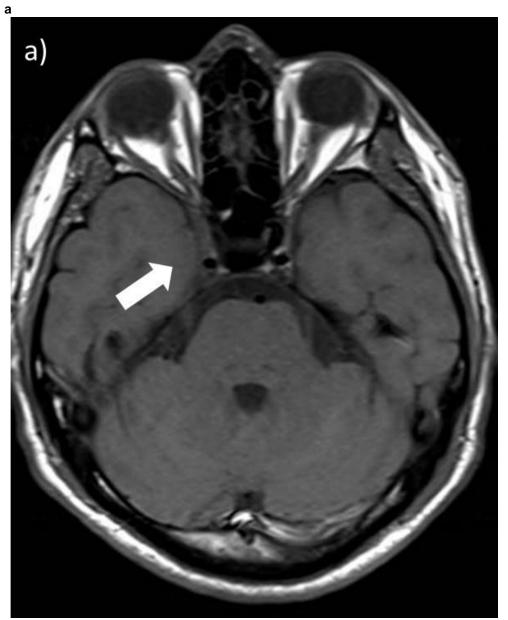


Description: b) Axial T1 fat saturation with contrast. The dural thickening has an homogenous enhancement with contrast. Notice the involvement of the intracavernous portion of the internal carotid artery. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina

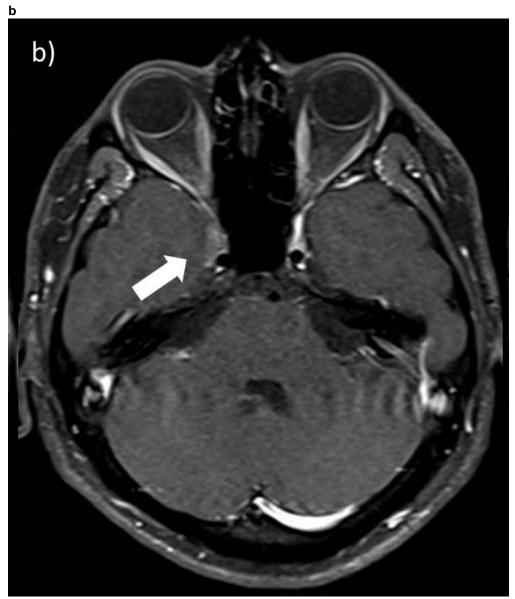


Description: c) Coronal T1 fat saturation with contrast. The dural thickening has an homogenous enhancement with contrast. Notice the involvement of the intracavernous portion of the internal carotid artery. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina

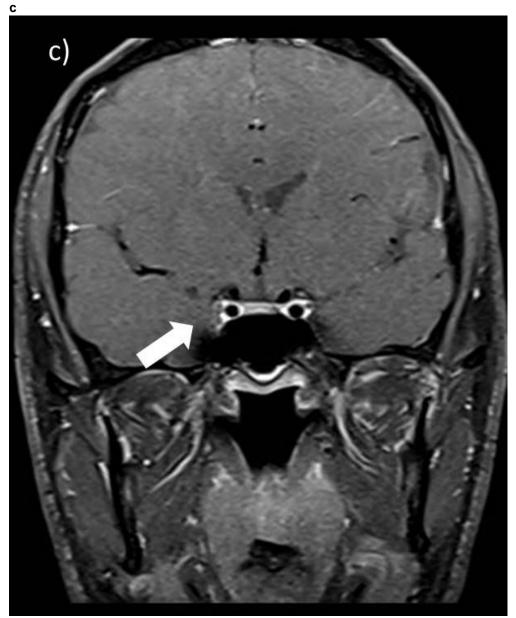
Figure 2



Description: a) Axial T1 SE. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina



Description: b) Axial T1 fat saturation with contrast. Significant decrease in thickness and enhancement of the lateral border of the right cavernous sinus. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina



Description: c) Coronal T1 fat saturation with contrast. Significant decrease in thickness and enhancement of dural thickening. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina

Figure 3



Description: Physical examination: restriction to abduction of the right eye due to paresis of the abducens nerve. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina

b



Description: Normal motility of the right eye to adduction. **Origin:** Imagenes MDQ, Clinic Pueyrredon, Department of Radiology, Mar del Plata, Argentina