Seeing Double! Tolosa-Hunt Syndrome

Neil Patel, DO, Henry Lam, DO, Arjan Ahluwalia, DO
Department of Medicine, Lehigh Valley Health Network, Allentown, PA

Introduction

Tolosa-Hunt syndrome (THS) is an exceedingly rare cause of diplopia with an estimated annual incidence of one case per million per year.

Case

- A 20-year-old female with history of migraines presented with binocular horizontal diplopia ongoing for the past four days.
- Exam revealed right cranial nerve VI palsy with the inability to abduct the right eye.
- Other extraocular movements were intact with reactive and equal pupils and unremarkable fundoscopic exam.
- Patient denied inciting factors but noted a right parietal headache ongoing for two months.

Figure 1. Representative image demonstrating right cranial nerve VI palsy with inability to abduct the right eye.

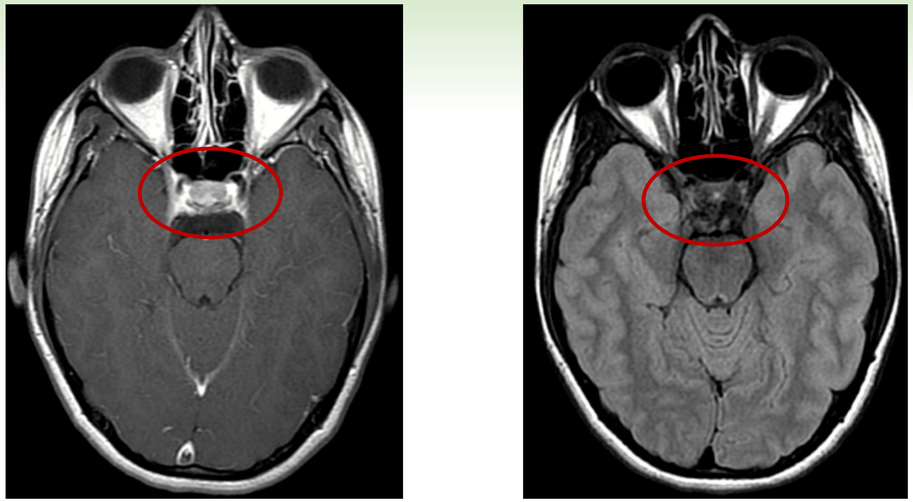

Figure 2. MRI brain T1 post contrast (*left*) and T2-FLAIR (*right*) showing asymmetric enhancement in the right cavernous sinus and along the cavernous and paraclinoid/supraclinoid segments of the right internal carotid artery.

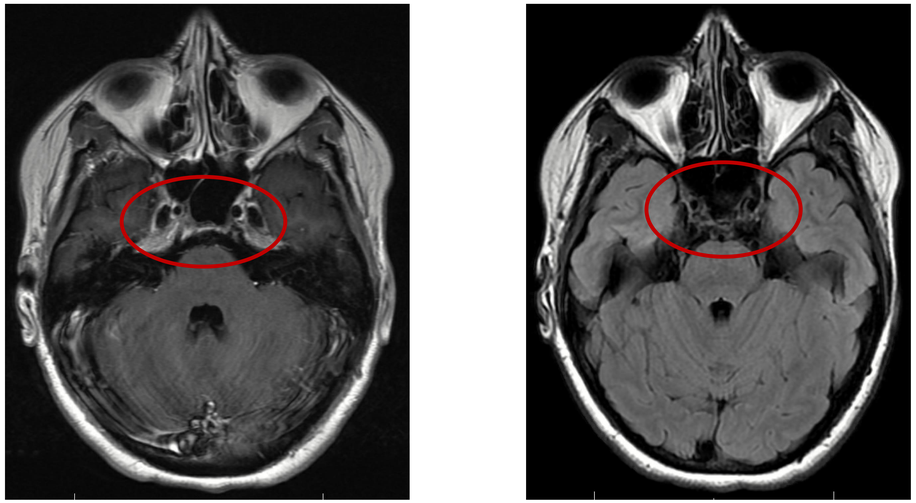

Figure 3. Follow up MRI brain T1 post contrast (*left*) and T2-FLAIR (*right*) showing improvement in the caliber of the cavernous sinus and right internal carotid artery inflammation.

Outcomes

- Expert evaluation by neurology determined the patient likely had THS.
- Extensive workup was unremarkable.
- Symptoms improved with high dose steroids.
- MRI two months later showed significant improvement in cavernous sinus inflammation.

Discussion

- THS is an uncommon ophthalmoplegia caused by cavernous sinus inflammation.
- Typical presentation is unilateral palsy of the third, fourth, or sixth cranial nerve.
- THS is a diagnosis of exclusion and relies upon a combination of patient presentation, neuroimaging results, and clinical response to steroids as noted in our patient.
- Complete resolution of symptoms can be expected; however, permanent deficits have been reported.

Conclusion

While rare, clinicians should consider THS as part of their differential diagnosis for diplopia especially in the absence of other etiologies.

References

1. Amrutkar C, Burton EV. Tolosa-Hunt Syndrome. [Updated 2021 Aug 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK459225/
2. Iaconetta G, Stella L, Esposito M, Cappabianca P. Tolosa-Hunt syndrome extending in the cerebello-pontine angle. Cephalalgia. 2005 Sep;25(9):746-50. doi: 10.1111/j.1468-2982.2005.00924.x. PMID: 16109058.
3. Mergen B, Sarici AM, Baltu F, Ozaras R, Adaletli I. An unusual presentation of sixth nerve palsy: neurobrucellosis. GMS Ophthalmology Cases. 2019 ;9:Doc13. DOI: 10.3205/oc000102. PMID: 31157155; PMCID: PMC6533445.
4. Yuliati A, Rajamani K. Tolosa-Hunt Syndrome. Neurohospitalist. 2018 Apr;8(2):104-105. doi: 10.1177/1941874417714147. Epub 2017 Jun 13. PMID: 29623162; PMCID: PMC5882006.