

Grand Rounds



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Subjective

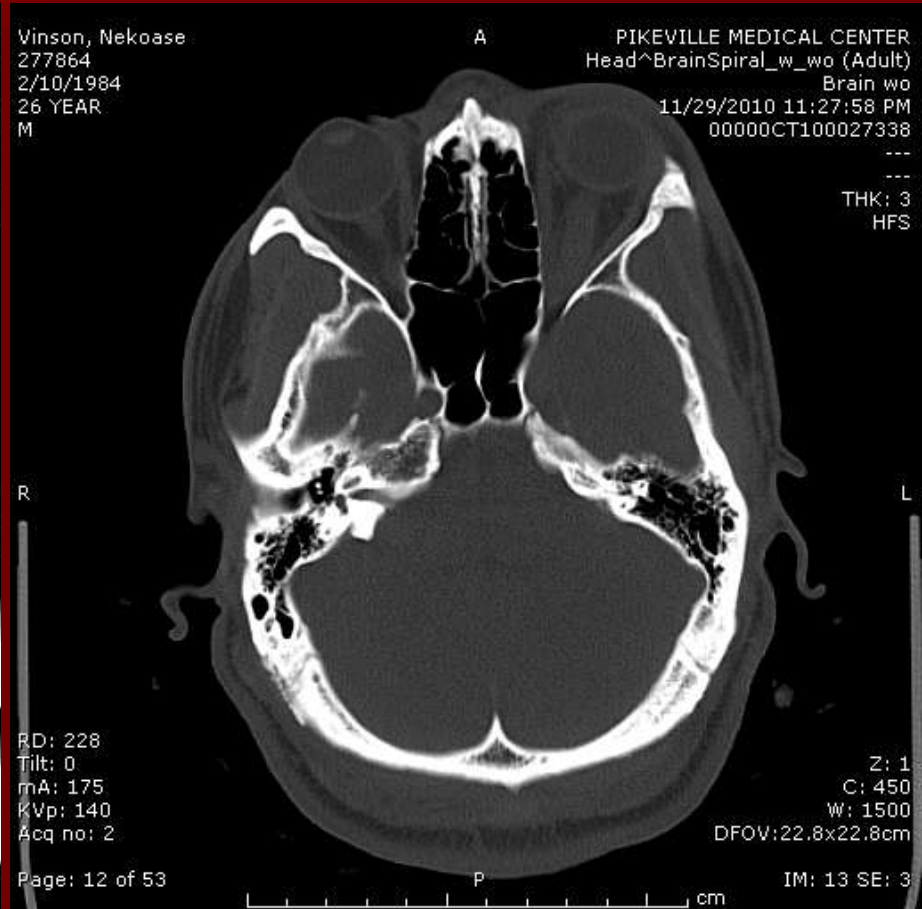
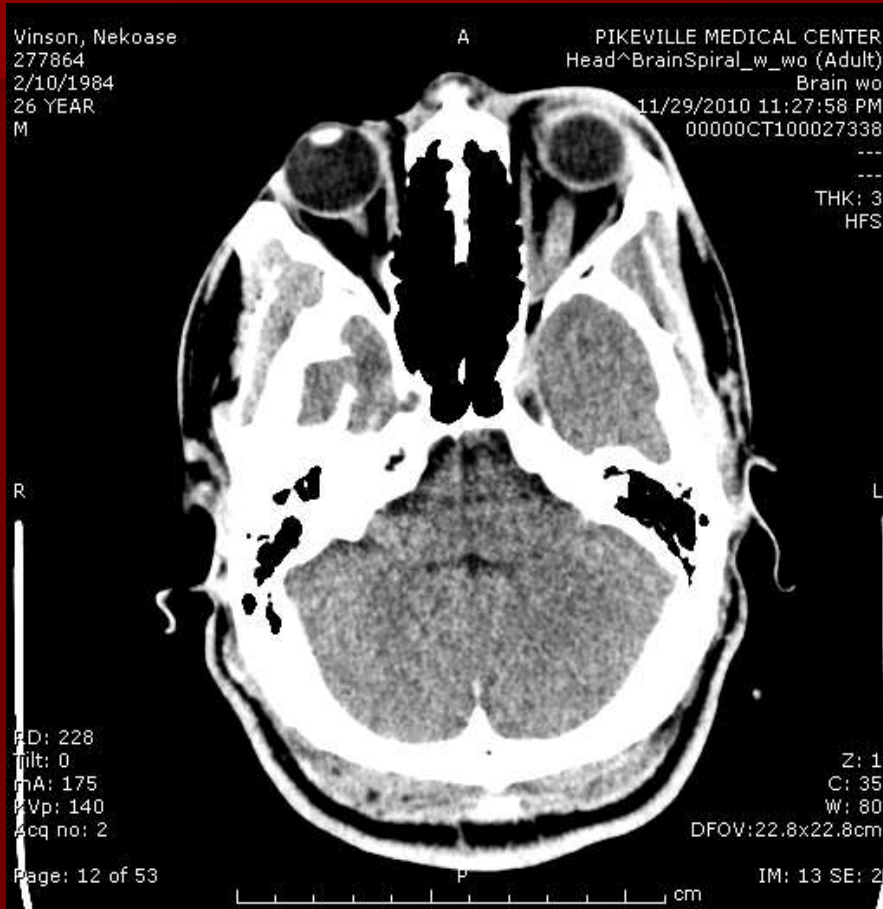
- CC: Pain and decreased vision in the left eye
- HPI: 26 year old AAM transferred from prison to ER with acute vision loss and an "orbital tumor" OS. Pt states his acuity has worsened over the previous 4 days and he can now only see the light. Also gives a 2 week history of headache and eye pain OS which have remained constant.
- ROS: denies trauma, recent illness or change in overall health. Has been imprisoned for 1 year.
- No previous medical or ocular history and taking no medications

Objective

	OD	OS
<u>Va(Sc):</u>	20/20	LP
<u>External:</u>	Left proptosis, eyelid ptosis -4 in all directions of gaze OS	
<u>Pupils:</u>	4mm → 2mm brisk	5mm non-reactive 3+RAPD OS
<u>IOP:</u>	15mmHg	13mmHg
<u>Ant Seg:</u>	WNL OU	
<u>DFE:</u>	Papilledema OS – Macula/Vessels/Periphery WNL OU	



CT



CT scan of brain showing diffuse enlargement of the left optic nerve with surrounding areas of fat stranding and proptosis

Labs on admission

- WBC: 9.3
- Hgb: 13.4
- Hct: 36.2
- Plts: 212

Assessment

- 26yo AAM with rapid onset painful vision loss, proptosis and multiple cranial neuropathies
- ? Orbital tumor OS on CT brain

Differential Diagnosis

- **Orbital pseudotumor→ Tolosa-Hunt**
- Orbital tumor-meningioma, glioma, rhabdomyosarcoma, lymphoma, metastatic disease
- Cavernous sinus thrombosis
- C-C fistula
- Infectious-Bacterial, fungal, TB, syphilis
- Sarcoidosis
- Wegners
- SLE
- Lyme

Plan

- Obtain MRI/MRV
- Recommended IV solumedrol to primary team
- Labs
 - CBC
 - ESR
 - ACE
 - ANA
 - ANCA
 - HIV
 - Lyme titers

Course-the following morning

- Primary team felt MRI and clinical findings were consistent with orbital tumor and therefore deferred IV steroids overnight

OS

Va(Sc):

NLP

External:

proptosis, pain and ptosis worse

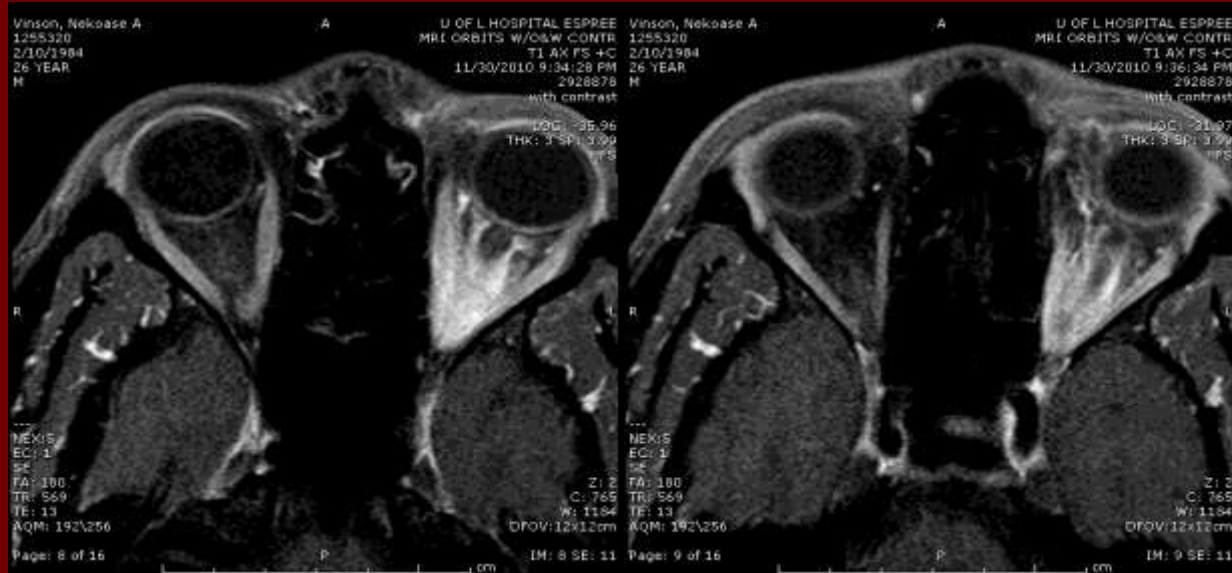
-4 in all directions of gaze OS

MRI



- Thickening of the left optic nerve with prominent enhancement surrounding the nerve and surrounding inflammatory material extending into the apex of the orbit
- Thickening and enhancement of the left rectus muscles.
- These findings are most concerning for an inflammatory process, such as idiopathic orbital pseudotumor.

MRI



LABS

- WBC: 9.7
- HGB: 16.4
- HCT: 46.9
- PLT: 299
- ESR: 1
- ACE: 31
- RPR: NR
- HIV: negative
- Lyme titers: negative

Course

- IV solumedrol started
- Pt still NLP this morning
- Ophthalmoplegia, proptosis and ptosis are resolving rapidly

Discussion



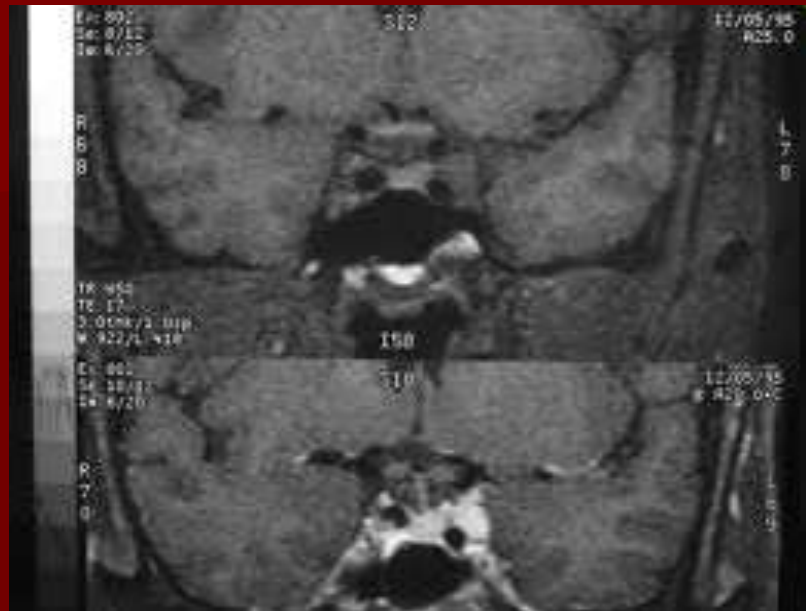
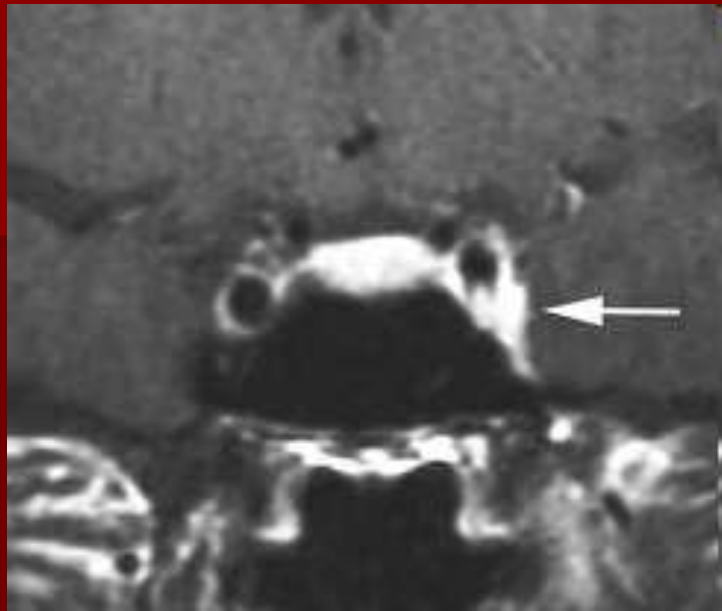
- Tolosa-Hunt syndrome (THS) is a painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure.
- The disorder is part of a continuum with idiopathic orbital pseudotumor.
- The diagnosis of Tolosa-Hunt syndrome is usually one of exclusion.

Discussion-Diagnosis

- The International Headache Society criteria for Tolosa-Hunt syndrome include the following:
 - Episode(s) of unilateral orbital pain for an average of 8 weeks if left untreated
 - Associated paresis of the third, fourth, or sixth cranial nerves, which may coincide with onset of pain or follow it by a period of up to 2 weeks
 - Pain that is relieved within 72 hours of steroid therapy initiation
 - Exclusion of other conditions by neuroimaging and angiography

Discussion-Imaging

- MRI of the brain and orbit with and without contrast, magnetic resonance (MR) angiography and CT scan of the brain and orbit with and without contrast may all be useful.
- Inflammatory changes in the cavernous sinus, superior orbital fissure, and/or orbital apex are typically observed on high-resolution contrast-enhanced imaging.
- Enlargement of the optic nerve or external ocular muscles has been described, emphasizing the continuum with idiopathic orbital inflammatory disorders.



Discussion

- **Laboratory Studies:** CBC, ESR, FTA, ANA, ANCA, Lyme titre, ACE level, and HIV titer are helpful in eliminating other processes.
- **Cerebrospinal fluid (CSF) studies:** Cell count and differential, protein, glucose, fungal and/or bacterial cultures, Gram stain, cytology, and opening pressure of CSF are helpful in eliminating conditions mimicking Tolosa-Hunt syndrome

Discussion-Treatment

- Corticosteroids are the treatment of choice, usually providing significant pain relief within 24-72 hours of therapy initiation.
- Ophthalmoplegia usually requires weeks to months for resolution
- For refractory cases, azathioprine (Imuran), methotrexate, or radiation therapy has been employed.
- The primary value of surgical intervention is a histopathologic diagnosis.

Discussion-Prognosis

- Patients usually respond to corticosteroids, although permanent ocular motor deficits may remain.
- Relapse can occur in as many as 40% of patients successfully treated for Tolosa-Hunt syndrome.
- This typically occurs on the same side as the original lesion but can be observed on the opposite side.
- Relapses may occur as long as 13 years after initial diagnosis and treatment.

Thank You

References

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- Rinsho Shinkeigaku. 2008 Apr;48(4):271-4.
- **["Tram-track" sign and "donut configuration" in Tolosa-Hunt syndrome]**
- [Article in Japanese]
- [Tamura A](#), [Taniguchi A](#), [Ochiai N](#), [Sasaki R](#), [Narita Y](#), [Kuzuhara S](#).
- Department of Neurology, Mie University Graduate School of Medicine.
- **Abstract**
- We report unique MRI abnormalities seen in a patient with Tolosa-Hunt syndrome (THS). A 60-year-old woman was admitted for left eye symptoms, including periorbital pain, numbness around the left eyebrow, blurred vision, delayed light reflex and impairment of abduction. Laboratory studies were unremarkable except for elevated erythrocyte sedimentation rates. T1-weighted MRI showed a mass lesion in the left orbital apex. Fat-suppressed T2-weighted MRI showed a hyperintense parallel linear lesion on the left optic nerve and a ring lesion around it, producing the "tram-track" sign on the axial view and the "donut configuration" on the coronal view. A diagnosis of THS was made, and corticosteroid therapy was started. Symptoms were improved rapidly, and MRI abnormalities disappeared. Reevaluation of MRI which had been taken at the previous episode of the right eye symptoms two years before also showed the "tram-track" sign and the "donut configuration" on the right. These signs are easy to be recognized and well reflect the stage of the disease. They are thus useful for diagnosing THS and evaluating the effect of the treatment. One should pay attention not only to the cavernous sinus and orbital apex, but also to the optic nerve for the MRI diagnosis of THS.

- J Med Imaging Radiat Oncol. 2008 Oct;52(5):447-51.
- **Tolosa-Hunt syndrome: MRI appearances.**
- Jain R, Sawhney S, Koul RL, Chand P.
- Department of Radiology, Sultan Qaboos University, Muscat, Oman.
rajeevjn@yahoo.com
- **Abstract**
- A review of MRI findings in seven patients with Tolosa-Hunt syndrome was carried out. Seven patients presented with unilateral painful ophthalmoplegia. Magnetic resonance imaging studies were carried out to evaluate the cavernous sinuses and orbits. Coronal fast spin-echo T2-weighted images and fat-saturated T1-weighted coronal and transverse images with and without contrast enhancement were obtained for the cavernous sinuses and orbits. All patients showed focal-enhancing masses expanding the ipsilateral cavernous sinus. In one patient the mass was extending to the orbital apex and intraorbitally. All patients recovered on corticosteroid therapy and resolution of the masses was documented on follow-up MRI studies in five patients. One patient had a relapse of symptoms after discontinuing therapy. Magnetic resonance imaging studies of the cavernous sinus and orbital apex show high sensitivity for the detection and follow up of inflammatory mass lesions in Tolosa-Hunt syndrome. Magnetic resonance imaging should be the initial screening study in these patients.

- Eur J Radiol. 2009 Mar;69(3):445-53. Epub 2007 Dec 31.
- **Tolosa-Hunt syndrome: MR imaging features in 15 patients with 20 episodes of painful ophthalmoplegia.**
- Schuknecht B, Sturm V, Huisman TA, Landau K.
- MRI Medical Radiological Institute Zurich, Switzerland. image-solution@ggaweb.ch
- **Abstract**
- PURPOSE: (a) To assess MR features in patients with Tolosa-Hunt syndrome (THS) and to (b) correlate MR findings with criteria derived from previously reported pathologic observations.
- METHODS: Fifteen patients with twenty episodes of painful ophthalmoplegia prospectively selected according to International Headache Society (IHS) standards underwent MR examinations focused on the cavernous sinus. Initial examinations in 20 and follow-up MR images in 17 episodes were retrospectively reviewed by 3 independent observers.
- RESULTS: The primary criteria: an enhancing soft tissue lesion within the cavernous sinus, increase in size and lateral bulging of the anterior cavernous sinus contour were consistently present in 15 initial episodes and in 5 recurrences (20/20). Agreement among observers was 100%. The secondary criteria: internal carotid artery narrowing in 7 patients, extension towards the superior orbital fissure in 13 and orbital apex involvement in 8 patients were unanimously agreed upon in 87.5%, 86.6% and 80%. Complete resolution of findings was observed on follow-up studies.
- CONCLUSION: In patients with THS the MR features conform to previously reported pathologic findings. MR features are evocative of THS when an increase in size and bulging of the dural contour of the anterior CS supplemented by carotid artery involvement and extension towards the orbit are present. Resolution of findings within 6 months is required to support the diagnosis.

