Confirmation of Tolosa-Hunt Syndrome via endoscopic biopsy of inflammation surrounding the orbital apex

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Objectives

1) Review the criteria of diagnosing Tolosa-Hunt Syndrome (THS), a rare cause of painful ophthalmoplegia due to idiopathic granulomatous inflammation of the orbital apex or cavernous sinus.
2) Review the literature of THS and other surgical techniques used previously in the setting of THS.

Case Report

- History of present illness: BH is a 41 year old male with 2 week history of decreased vision, left eye pain, pressure headache. No light perception from his left eye for 5 days.
- Surgical / social history: appendectomy, current 1/2 pack-per-day smoker, social drinker, no illicit drugs
- Vital signs: Temperature 100.3F
- Exam:
  - Eyes: No light perception in left eye, afferent pupillary defect in left eye, paralysis of extraocular movements (total loss of cranial nerves III, IV, VI), right eye normal
  - Neuro: Decreased sensation in left CN V2
  - CBC, CMP, ESR, CRP, CSF, CTA, MRA, MRV all normal
  - MRI demonstrated abnormal enhancement in left orbital apex and cavernous sinus with extension along left infraorbital nerve into the left inferior orbital and pterygopalatine fissures.
  - CT demonstrated loss of fat planes in the left inferior orbital and pterygopalatine fissures and hyperostosis of the left sphenoid bone.

Diagnostic Criteria

- 2004 THS International Headache Society criteria
  - A. One or more episodes of unilateral orbital pain persisting for weeks if untreated
  - B. Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or demonstration of granulomatous inflammation by MRI or biopsy
  - C. Paresis coincides with the onset of pain or follows it within 2 weeks
  - D. Pain and paresis improve within 72h when treated adequately with corticosteroids
  - E. Other causes have been excluded by appropriate investigations

Background

- One case per million per year, rare cause of painful ophthalmoplegia
- Nonspecific inflammation of orbital apex, septa and wall of cavernous sinus: lymphocyte and plasma cell infiltration, giant cell granulomas, and proliferation of fibroblasts
- Palsy of CN III, IV, VI, and superior divisions of CN V
- Differential diagnosis includes tumors, vasculitis, basal meningitis, sarcoid, diabetes mellitus, and ophthalmoplegic 'migraine'
- MRI demonstrates inflammation in the cavernous sinus, superior orbital fissure or orbit in 92% of cases
- Diagnosis is a diagnosis of exclusion or demonstration of granuloma on MRI or granulomatous inflammation on histopathology
- Treatment with steroids; steroid type, dosage, administration method, and duration are variable

Pathology

- Biopsies performed of pterygopalatine fossa, pterygoid fossa, and sphenopalatine foramen
- Figure 1 demonstrates chronic inflammation with distorted lymphocytes (arrowhead) and plasma cells and fibrosis
- Figure 2 demonstrates chronic granulomatous inflammation with distorted lymphocytes surrounding fibrovascular bundles (arrowhead)

Operative Biopsy

- Endonasal endoscopic biopsy of inflammatory tissue at pterygopalatine fossa and sphenopalatine foramen was performed within 72 hours to confirm chronic granulomatous inflammation and rule out infectious and neoplastic etiology.
- Steroids were initiated, and patient BH had resolution of his pain within 24 hours of starting high dose intravenous dexamethasone. He continued a several week course of oral steroids and continued to have gradual improvement of his ophthalmoplegia. At his one month follow up visit, he had significantly improved vision.
- Historically, THS has been a diagnosis of exclusion. There are only 14 documented cases with histologic diagnosis, of which only one biopsy in vivo was performed via transnasal approach (sphenoidotomy) and only 50% demonstrated granuloma formation or granulomatous inflammation. Most cases of THS are confirmed by symptoms and granuloma on imaging.
- Endonasal endoscopic biopsy of the peri-orbital apex proved to be a safe, low risk procedure to confirm THS and rule out dangerous causes of painful ophthalmoplegia, including malignancy.

Imaging

- Image 1. Coronal fat suppressed MRI. Enhancement of extraocranial muscles at orbital apex and extension into pterygopalatine fossa (arrow).
- Image 2. Axial fat suppressed MRI. Enhancement of extracranial optical nerve extending into inferior orbital fissure (arrow).
- Image 3. Coronal CT with contrast. Loss of fat planes in pterygopalatine fossa at sphenopalatine foramen (arrow).

Surgical Approaches

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<td>Tolosa. 1954</td>
<td>1</td>
<td>Transfrontal intradural, autopsy</td>
<td>Negative; non-specific periarterial granulation</td>
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<td>Hunt et al. 1961</td>
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<td>Lakk. 1982</td>
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<td>Schatz. 1972</td>
<td>2</td>
<td>Frontotemporal craniotomy to cavernous sinus</td>
<td>Chronic inflammation, plasma cells, fibrosis; chronic granulomatous lesion</td>
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<td>Campbell et al. 1987</td>
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<td>Brown et al. 1990</td>
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<td>Sublabial transseptal sphenoidotomy</td>
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<td>Yousem et al. 1990</td>
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<td>Unspecified cavernous sinus biopsy</td>
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<td>Lance. 1991</td>
<td>1</td>
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<td>Lateral cavernous sinus granuloma</td>
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<td>Montecucco et al. 1993</td>
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<td>Mononuclear cells noncaseating granulomas</td>
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<tr>
<td>Wasmeier et al. 2002</td>
<td>2</td>
<td>Transfrontal biopsy of medial rectus; transorbital exploration</td>
<td>Granulocytic eosinophilic granulomatous inflammation at orbital apex</td>
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References


Disclosure

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