Neurosarcoidosis: Presentation as a Skull Base Lesion

Kenneth C. Iverson MD,
Carrie M. Bush MD, Hannah Coulson DO,
Dilip A. Thomas MD, Michael Toscano MD,
C. Arturo Solares MD

Georgia Health Sciences University
Department of Otolaryngology / Head & Neck Surgery

Case Presentation

• 30 year old black female, 5 months history of progressive left sided:
  • Periorbital headache
  • Proptosis
  • Decreased visual acuity
  • Decreased facial sensation and strength
  • Nausea and emesis
  • Treated for cluster headaches

Case Presentation

• PMH: Headaches, HTN
• PSH: None
• SH: Single mother of 3 children, 10 pack year smoking history
• FH: Mother with cancer

Case Presentation

• Physical exam:
  • Left proptosis
  • Decreased left visual acuity
  • Decreased left facial sensation
  • House-Brackmann II on left
  • Remaining neurological exam normal

Case Presentation

• Nasal endoscopy
  • Medialized left middle turbinate
  • Left middle meatal fleshy mass
  • Bilateral infiltrated nasopharynx

• Financial disclosures
  • Nothing to disclose
Case presentation

Initial biopsy in the office:
- Mixed B, T, and plasma cells
- Negative for malignancy
- Flow cytometry – no aberrant immunophenotype

Nasal endoscopy with biopsy in OR
- Negative AFB and fungal stains/cultures
- Aerobic cultures: +MSSA

Ciliated columnar sinonasal mucosa
- Submucosal non-caseating granulomata
- Numerous Langhans-type giant cells

CXR
- Prominent bilateral hilar adenopathy
- Chronic interstitial changes

Laboratory
- Elevated ACE
- 59 units/L
Case Presentation

- **Initial Therapy**
  - Prednisone 60 mg QD
  - 2 months
  - Mycophenolate mofetil 1 gm BID added
  - 4 months
  - Prednisone 50mg QD
  - Mycophenolate mofetil to 1500mg BID

- **Clinical Response**
  - Initial improvement in vision
  - OS central scotoma and bitemporal visual field loss OS>OD
  - Resolution of CN V and VII symptoms
  - Headache resolution

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Sarcoidosis

- Multisystem granulomatous disease
- Unknown etiology
- Lungs, skin, & eye involvement most common
- 5-15% with neurologic complications


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- Ubiquitous worldwide
- Environmental predisposition
  - Aerosolized metal exposure
  - *Mycobacterium* and *Propionibacterium*
- Genetic predisposition
  - West African
  - Northern European
Neurosarcoidosis

- Difficult diagnosis
- Any part of nervous system affected
  - Variable presentation
- No specific sign or symptom
- Histopathologic ambiguity

Neurosarcoidosis

- Present as neurologic condition or skull base lesion

<table>
<thead>
<tr>
<th>TABLE 1. Neurological Manifestations</th>
<th>Frequency</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dural neuropathy</td>
<td>50%-70%</td>
<td>Good</td>
</tr>
<tr>
<td>Parasympathetic nervous lesions</td>
<td>50%</td>
<td>Fair</td>
</tr>
<tr>
<td>Cognitive/behavioural manifestations</td>
<td>20%</td>
<td>Good</td>
</tr>
<tr>
<td>Mental disease</td>
<td>16%-30%</td>
<td>Poor</td>
</tr>
<tr>
<td>Peripheral neuropathy</td>
<td>15%</td>
<td>Fair</td>
</tr>
<tr>
<td>Sjogren†</td>
<td>5%-10%</td>
<td>Good</td>
</tr>
<tr>
<td>Visual loss</td>
<td>5%-10%</td>
<td>Fair</td>
</tr>
<tr>
<td>Myopathy</td>
<td>1.4%-2.3%</td>
<td>Fair</td>
</tr>
</tbody>
</table>

*Acute is defined as <3 months, chronic is defined as >3 months. If there is active brain inflammation or scarring may be confounding. If the inflammation may process is controlled certain vaccines may respond to standard anti-inflammatory medications. Terushkin, et al. Neurosarcoidosis: Presentation and Management. The Neurologist 2010;16:1-15.

Neurosarcoidosis

- Predilection for the skull base
  - CN VII
    - Most common neuropathy (50-65%)
  - CN II
    - 2nd most common neuropathy
    - Most common affected on imaging
- Neuroendocrine-related symptoms

Neurosarcoidosis

- Greater than 90% have systemic signs of sarcoidosis
- Up to 30% of sarcoidosis initially present as neurosarcoidosis
- Rare presentations of isolated neurosarcoidosis

Neurosarcoidosis

- Systemic Testing
  - CNS and pulmonary imaging
  - ACE level
  - CSF testing
- Non-specific
- "The diagnosis of sarcoidosis is never definitive.”

Neurosarcoidosis

• Therapy
  • Early initiation to prevent:
    • Acute CNS complications
    • Permanent CNS damage

  • Prednisone
    • 0.5 – 1 mg/kg/day with taper
    • Up to 1 year of treatment

• Other anti-inflammatory medications
  • Methotrexate (MTX)
  • Mycophenolate mofetil (MMF)
  • Cyclophosphamide
  • Azathioprine
  • Chloroquine and hydroxychloroquine
  • Thalidomide
  • Infliximab

Neurosarcoidosis

• Radiotherapy
  • Reserved for pharmacotherapy failure or intolerance
    • 1.5 Gy/d
    • Total dose 20 Gy

• Surgical therapy
  • Diagnosis
  • Life-threatening medical failure
    • Shunting
    • Resection

Conclusion

• Neurosarcoidosis diagnosis should be considered for a skull base lesion
• Must maintain high level of suspicion
• Systemic testing should be conducted

Neurosarcoidosis

• Diagnosis
• Life-threatening medical failure
  • Shunting
  • Resection