AMSER Case of the Month
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33 y/o F with bilateral paresthesias of the hands and wrists

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Patient Presentation

• 33 y/o female with persistent paresthesias of the hands and wrists bilaterally
• PMH: Hepatitis C and IV drug use
• PSH: Cholecystectomy
• Nerve conduction studies showed bilateral median & ulnar n. changes
  • C5, C6
    • chronic active right sided radiculopathies & ongoing denervation of the right biceps brachii & pronator teres
    • chronic active left sided radiculopathies & ongoing degeneration of the left deltoid & biceps brachii
  • C7, C8
    • demonstrated evidence of inactive radiculopathies bilaterally
Differential diagnosis:

- Multiple Sclerosis
- Carpal Tunnel
- Malignancy
- Degenerative Disk Disease
- Nerve Compression, osseous
- Spinal Ischemia
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

Variant 2: New or increasing nontraumatic cervical radiculopathy. No “red flags.” Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI cervical spine without IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>CT cervical spine without IV contrast</td>
<td>May Be Appropriate</td>
<td>⚫⚫⚫⚫⚫</td>
</tr>
<tr>
<td>Radiography cervical spine</td>
<td>May Be Appropriate (Disagreement)</td>
<td>⚫⚫</td>
</tr>
<tr>
<td>MRI cervical spine with IV contrast</td>
<td>Usually Not Appropriate</td>
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<td>Radiographic myelography cervical spine</td>
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</tr>
<tr>
<td>CT cervical spine without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>⚫⚫⚫</td>
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<tr>
<td>CTA neck with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>⚫⚫⚫</td>
</tr>
<tr>
<td>Discography cervical spine</td>
<td>Usually Not Appropriate</td>
<td>⚫⚫</td>
</tr>
<tr>
<td>Facet injection/medial branch block cervical spine</td>
<td>Usually Not Appropriate</td>
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<td>MRA neck with IV contrast</td>
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<tr>
<td>Bone scan whole body with SPECT or SPECT/CT neck</td>
<td>Usually Not Appropriate</td>
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This imaging modality was ordered by the chiropractic physician.
Findings (labeled)

- Interlaminar space narrowing posteriorly
- Degenerative facet changes
- Loss of lordosis

UPRIGHT
AP
Lateral
Select the applicable ACR Appropriateness Criteria

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This imaging modality was ordered due to continued symptoms & neurologist recommendation.
Findings (labeled)

Intramedullary isointense mass on T1

Intramedullary hyperintense mass on T2

Ill defined hyperintensity in the adjacent spinal cord

Susceptibility artifact rim inferiorly
Final Dx:
Intramedullary Spinal Cord Ependymoma (WHO Grade 2)
Characteristic Imaging Findings

• Plain film
  • Canal widening
  • Vertebral body scalloping
  • Pedicle erosion
  • Laminar thinning

• CT
  • Non-specific canal widening
  • Mass may be a similar or lower attenuation than white matter
  • Mass enhancement with IV contrast
Characteristic Imaging Findings

• MRI
  • Circumscribed Intramedullary (within the spinal cord) mass
  • T1: isointense to hypointense, or heterogenous in signal intensity
  • T2/STIR: isointense to hyperintense
    • peritumoral edema (as seen in this case)
      • Hyperintense signal around the mass
    • Hemosiderin cap sign (as seen in this case): susceptibility artifact from prior hemorrhage/hemosiderin deposition, a common finding
      • Susceptibility artifact: disruption of magnetic fields causing image distortions appearing hypointense on all sequences

• Other findings
  • Syringohydromyelia
    • Fluid in the central canal of the spinal cord
  • Tumoral cysts and non-tumoral cysts
Case Discussion

• Epidemiology:
  • Most common intramedullary neoplasm in adults
  • Second most common in children
  • Peak incidence is the 4th decade of life, male > female
  • Association with NF2

• Presentation:
  • Pain, weakness, sensory deficits

• Pathology:
  • Arise from embryonic rests of ependymal tissue
  • Perivascular pseudorosettes
  • MYCN amplification association
  • 60% infratentorial, 30% supratentorial & 10% in spinal cord (as seen in this case)
    • Infratentorial tend to be solid, supratentorial tend to be cystic
Case Discussion: WHO Grading System

• Grade I:
  • Myxopapillary
    • Slow growing, good long-term survival
    • Exclusively in conus-cauda-filum-terminale region → low back pain presentation
  • Subependymoma
    • Slow growing, noninvasive, usually does not present in the spinal cord

• Grade II:
  • Four variants: cellular, papillary, clear cell, and tancytic
  • Pseudorossettes and less commonly ependymal rosettes
  • GFAP or EMA differentiation
  • RELA positive can be grade II or III

• Grade III:
  • Anaplastic, microvascular proliferation, central necrosis
  • Decreased T2 signal on MRI
Case Discussion: Treatment and Prognosis

- Slow growing, compressing spinal cord
- Complete curative resection achieved in 50% of patients
  - 5-year survival = 85%
  - Incomplete resection: 57% 5-year survival
- Metastatic spread is rare
- Radiation therapy can be used for high grade tumors or incomplete resection
- MYCN-amplification are aggressive tumors and spread to central nervous system and have high reoccurrence
References:


