RADY 401 Case Presentation

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Focused patient history and workup

**Initial Presentation**

2-year-old male presents to PCP with abdominal distension and 6-8 episodes of diarrhea daily for the last year. No significant medical history.

**1 Week later**

Presents with vomiting, lethargy, and 4 lb weight loss

→ Admitted for dehydration and hypokalemia
→ **Abdominal x-ray** shows dilated bowel
→ **Contrast enema** yields distended colon without fecal material. Rectal biopsy negative for Hirschsprung disease
Continued patient workup

3 Weeks later

Continues to have problems feeding and failure to thrive
→ GI surgery team attempts to place G-tube and discovers “peach”-sized mass in the retroperitoneum.
→ CT Abdomen Pelvis with IV contrast
→ FDG-PET scan with CT
→ Bone marrow and mass biopsy negative

4 Weeks later

Suspect neuroblastoma
→ Biopsy confirms diagnosis (but it is not neuroblastoma)
List of imaging studies

• Abdominal radiograph
• Contrast enema (not pictured)
• CT Abdomen and Pelvis with IV Contrast
• FDG-PET Scan with CT
Abdominal radiograph

Dilated Bowel Loops

Calcifications

Note: We can see contrast in the descending colon and sigmoid colon. This is likely from a prior study.
Dilated bowel loops are anterior to the retroperitoneal mass. Calcifications are more readily seen (than on the plain radiograph).
Avid uptake of the retroperitoneal mass with sparing of the SMA and descending aorta together suggest that the mass may be folding around important structures as opposed to invasive growth.

Intact adrenal glands with low uptake bilaterally suggest that this is NOT the primary tumor source.

Uptake of the kidneys, bladder, brain and liver are all normal in this case.
Neuroblastic Tumors

- Neuroblasts are immature sympathetic cells of neural crest origin which differentiate into ganglion and Schwann cells.
- More often found in children and more often malignant.
- Locations: adrenal gland, retroperitoneum and posterior mediastinum.
- Symptoms: Pain, abdominal distension, wt. loss and neuro deficits.
- Radiological findings: heterogeneous mass +/- calcifications.

Neuroblastoma (NB)
Ganglioneuroblastoma (GNB)
Ganglioneuroma (GN)
Staging of Neuroblastic Tumors

INRG SS
Assessment made prior to surgery

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>L1</td>
<td>Local tumor with IDRF = 0</td>
</tr>
<tr>
<td>L2</td>
<td>Local tumor with IDRF ≥ 1</td>
</tr>
<tr>
<td>M</td>
<td>Mets anywhere</td>
</tr>
<tr>
<td>MS</td>
<td>Mets to skin, liver or BM (&lt; 18 mo)</td>
</tr>
</tbody>
</table>

*IDRF = Image Defined Risk Factors

Bone marrow biopsy and PET scan were negative for distant metastasis so this case is likely grade L2 given the radiologic findings.

Table 1A. IDRF – Image Defined Risk Factors in neuroblastic tumors

<table>
<thead>
<tr>
<th>Ipsilateral tumor extension within two body compartments:</th>
<th>Neck: chest, chest-abdomen, abdomen-pelvis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck:</td>
<td>Tumor encasing carotid and/or vertebral artery and/or internal jugular vein</td>
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<tr>
<td></td>
<td>Tumor extending to base of skull</td>
</tr>
<tr>
<td></td>
<td>Tumor compressing the trachea</td>
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<tr>
<td>Cervico-thoracic junction:</td>
<td>Tumor encasing brachial plexus roots</td>
</tr>
<tr>
<td></td>
<td>Tumor encasing subclavian vessels and/or vertebral and/or carotid artery</td>
</tr>
<tr>
<td></td>
<td>Tumor compressing the trachea</td>
</tr>
<tr>
<td>Thorax:</td>
<td>Tumor encasing the aorta and/or major branches</td>
</tr>
<tr>
<td></td>
<td>Tumor compressing the trachea and/or principal bronchi</td>
</tr>
<tr>
<td></td>
<td>Lower mediastinal tumor, infiltrating the costo-vertebral junction between T9 and T12</td>
</tr>
<tr>
<td>Thoraco-abdominal:</td>
<td>Tumor encasing the aorta and/or vena cava</td>
</tr>
<tr>
<td>Abdomen/pelvis:</td>
<td>Tumor infiltrating the porta hepatitis and/or the hepaticoduodenal ligament</td>
</tr>
<tr>
<td></td>
<td>Tumor encasing branches of the superior mesenteric artery at the mesenteric root</td>
</tr>
<tr>
<td></td>
<td>Tumor encasing the origin of the celiac axis, and/or of the superior mesenteric artery</td>
</tr>
<tr>
<td></td>
<td>Tumor invading one or both renal pedicles</td>
</tr>
<tr>
<td></td>
<td>Tumor encasing the aorta and/or vena cava</td>
</tr>
<tr>
<td></td>
<td>Tumor encasing the iliac vessels</td>
</tr>
<tr>
<td></td>
<td>Pelvic tumor crossing the sacral notch</td>
</tr>
<tr>
<td>Infiltration of adjacent organs/structures:</td>
<td>Pericardium, diaphragm, kidney, liver, duodeno-pancreatic block and mesentry</td>
</tr>
</tbody>
</table>

Table 1B. Conditions to be recorded, but NOT considered IDRFs:

- Multiloculated tumors
- Intraspinal tumors (with or without symptoms of spinal cord compression. For staging of patients with intraspinal tumors: see text)
- Pleural effusion (with or without malignant cells)
- Ascites (with or without malignant cells)
Patient treatment

- Biopsy confirms ganglioneuroma
- Patient awaits excision of mass

### Standard Treatment and Prognosis for Neuroblastic Tumors

<table>
<thead>
<tr>
<th>Neuroblastic Tumor Subtype</th>
<th>Cells</th>
<th>Prognosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neuroblastoma</td>
<td>Immature (malignant)</td>
<td>Poor</td>
<td>Surgery and chemo +/- bone marrow transplant</td>
</tr>
<tr>
<td>Ganglioneuroblastoma</td>
<td>Immature and mature (malignant)</td>
<td>Intermediate</td>
<td>Surgery and chemo</td>
</tr>
<tr>
<td>Ganglioneuroma</td>
<td>Mature (benign)</td>
<td>Excellent</td>
<td>Surgery</td>
</tr>
</tbody>
</table>
Imaging discussion

• Initial abdominal radiograph – appropriate
  • To evaluate for suspected small bowel obstruction in a child while minimizing radiation exposure

• Contrast enema (water soluble) – appropriate
  • To evaluate for distal bowel obstruction in child
  • R/o appendicitis, hirschsprung’s, meconium plug, atresia (or f/u if still suspected)

• CT Abdomen and Pelvis with Contrast – appropriate
  • To evaluate the retroperitoneal mass (suspected Neuroblastoma)

• PET scan – appropriate
  • To identify tumor location. Alternatively could have used MIBG (rather than FDG) tracer
# Approximate Cost of Imaging Studies

<table>
<thead>
<tr>
<th>Imaging</th>
<th>Radiation exposure*</th>
<th>Cost</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal x-ray</td>
<td>0.7 mSv</td>
<td>$23 - $380</td>
</tr>
<tr>
<td>Contrast enema with Fluoro</td>
<td>8 mSv</td>
<td>$242 - $620</td>
</tr>
<tr>
<td>CT Abdomen and Pelvis</td>
<td>10 mSv</td>
<td>$889 - $4,050</td>
</tr>
<tr>
<td>PET scan with CT</td>
<td>up to 25 mSv</td>
<td>$1,526 - $3,738</td>
</tr>
</tbody>
</table>

*Exposure is approximated for an adult.*
Imaging discussion for MIBG-PET Scan

• $^{123}$Iodine metaiodobenzyl guanidine ($^{123}$I-MIBG) is used for diagnostic workup of neuroblastoma
  • MIBG, an analog of norepinephrine (NE), is taken up by NE transporters and accumulates in cells.
  • > 90% of NB are MIBG avid; however, other tumors can have this uptake pattern (e.g. NB, GNB, GN, pheo, carcinoid, medullary thyroid). This means MIBG cannot definitively differentiate among these tumors.

• Sensitivity (90%) and Specificity (88%) for neuroblastoma, but MIBG uptake is variable for ganglioneuroma.

• Regardless of the tracer used, confirm via primary biopsy, bone marrow biopsy or catecholamine metabolites.
UNC Top Three

• With the presence of heterogeneous mass in the adrenal glands, retroperitoneum, or posterior mediastinum (especially in a child), suspect neuroblastic tumor.

• Ganglioneuroma is a rare but benign tumor that presents similarly to neuroblastoma.

• $^{123}$I-MIBG can be used to identify various tumors of neural crest origin (NE uptake), but particularly neuroblastoma.
References


