AMSER Case of the Month
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HPI: 9-month-old female with a neck mass

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Patient Presentation

• **HPI:** 9-month-old female with no significant past medical history who has swelling in her neck with a question of possible branchial cleft remnant. The mass has increased in size and has become firmer. It is relatively mobile. The patient has no symptoms related to the mass. Specifically no respiratory symptoms, wheezing, or stridor. All other ROS are negative.

• **PMHx/PSHx/FH:** Born full term via NSVD. No other past medical history, no previous surgeries, and no known allergies. The patient lives with her parents and has two siblings. No home exposure to tobacco or any illicit substances. Family history is noncontributory.

• **Relevant Meds:** None.
Patient Presentation

• **Vitals**: Current weight is 9 kg. Height of 71.1 cm.

• **Physical Examination**: In no acute distress, happy on mother's lap. Had a palpable somewhat mobile mass, approximately 1.5 to 2 cm within the left side of the neck. Unremarkable otherwise.

• No pertinent labs.
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

**Variant 4:** Child. Neck mass(es). Not parotid region or thyroid. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>CT neck with IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI neck without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
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<tr>
<td>US neck</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI neck without IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
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<tr>
<td>CT neck without IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
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<tr>
<td>MRA neck without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
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<tr>
<td>MRA neck without IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>CT neck without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>CTA neck with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>Arteriography cervicocerebral</td>
<td>Usually Not Appropriate</td>
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<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>FDG-PET/MRI skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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These imaging modalities were ordered by the physician.
Findings (unlabeled)

Neck Ultrasound
Findings (unlabeled)

CT Neck w/ Contrast
Findings (unlabeled)

CT Neck w/ Contrast
Findings: (labeled)

Solid mildly vascular mass located in the left lateral neck measuring 1.7 cm x 1.1 cm in long and short axis.

The lesion is heterogeneous with multiple echogenic foci. The lesion is separate from the thyroid and thymus.
Findings: (labeled)

Neck Ultrasound

Second heterogenous solid mass with multiple echogenic foci visualized on angling inferior to the palpable lesion. The mass is medial and inferior to the sentinel lesion extending into the chest and measures 3.5 cm by 3.5 cm in AP and transverse dimensions.
Findings: (labeled)

Enhancing partially calcified mass deep to the sternocleidomastoid muscle. Located slightly posterior to the left internal jugular vein. This lesion measures 18.6 mm x 13.2 mm
Inferior to the mass outlined in the previous slide is a similar soft tissue mass with internal calcification. The mass is in the left lung apex/paraspinal region measuring 33.2 mm x 24.1 mm. There is no definite neuroforaminal extension.
Additional CT of the chest, abdomen, and pelvis were ordered.
CT Abdomen and Pelvis with Contrast

Coronal image of the neck

Sagittal image of the neck

Two adjacent lesions. The primary lesion is in the posterior mediastinum.

It is difficult to tell whether this lesion is a calcified lymph node or an extension of the tumor.

No additional sites of disease. There is no primary lesion in the adrenal glands.
Final Dx:

**Surgical Pathology:** Metastatic neuroblastoma (Poorly differentiated, schwannian stromal poor, Mitosis-Karyorrhexis Index low to intermediate making it favorable histology)
Case Discussion: Overview

- Neuroblastoma is a malignant neuroendocrine tumor of the sympathetic nervous system with an overall incidence of 1 case per 10,000 births.\(^3\) It is the most common extracranial solid tumor in the pediatric population, accounting for 8% to 10% of all childhood tumors.\(^1\)

- The cancer originates from neural crest cells and is most commonly found in the adrenal glands or along the sympathetic chain.\(^1\) In over 60% of neuroblastoma cases, the location of the primary tumor is the abdomen. The primary tumor can also be found the chest and neck such as in our patient.

- Clinical features of neuroblastoma localized to the chest and neck include Horner syndrome, spinal cord compression (back pain, weakness, numbness, ataxia, loss of bladder or bowel control, cough, and dyspnea).\(^2\)
Case Discussion: Imaging

- Ultrasound is the primary first-line modality for detecting neuroblastoma, specifically for tumors located in the neck, abdomen, and pelvis.

- Suggestive features of neuroblastoma on ultrasound include internal calcifications (seen in this case) and encasement of vessels.\(^4\)

- The main imaging modalities utilized for staging neuroblastoma in patients are CT, MRI, and MIBG scans.

- MRI is recommended over CT, especially in cases of spinal canal involvement. MRI has been found to be more sensitive for detecting local disease, bone marrow disease, chest wall invasion, and stage 4 disease.\(^4\)

- MRI is also preferred due to its inherent contrast, lack of ionizing radiation, and multiplanar imaging capability.\(^5\)
Case Discussion: Treatment

• Outcomes/prognosis vary and are dependent on the specific biology of the tumor. Overall, the disease accounts for 15% of cancer-related mortalities in children. \(^3\)

• Treatment for neuroblastoma typically includes a combination of surgery, chemotherapy, and radiotherapy. Surgery is performed to either achieve complete resection of the tumor or stage the tumor via examination and/or biopsy. Imaging is pivotal for determining whether surgical intervention is the best option and determine the timing of surgery.

• Common chemotherapy agents utilized in neuroblastoma treatment include cyclophosphamide, iphosphamide, vincristine, doxorubicin (adriamycin), cisplatin, carboplatin, etoposide (VP-16) and melphalan. \(^3\)

• Radiation therapy can reduce local relapse rates and is primarily utilized for higher risk tumors. It has been found to work well in combination with chemotherapy to reduce the tumor burden and thus allow for surgical resection afterwards.
References:


