AMSER Case August 2020

12 Year Old Male with Cough and Persistent Cervical Lymphadenopathy

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Patient Presentation

12-year-old previously healthy, unvaccinated male presents to clinic with a 6 week history of cervical lymphadenopathy, 3 weeks of fatigue and 1 week of mild cough.

- HPI:
 - 6 weeks prior, developed hives on the lower abdomen, bilateral cervical lymphadenopathy, pharyngitis and headache after pulling weeds near a woodpile. Lymphadenopathy persisted and patient developed fatigue, mild cough with "noisy breathing" when sleeping.
 - Prior tests: Negative for mumps, EBV and Lyme; CXR at onset within normal limits; CBC with differential unremarkable both at onset 6 weeks ago and again 3 weeks ago.
- ROS
 - Fatigue, mild cough, muscle pain at nape of neck and upper back
 - Denies fever, weight loss, voice changes or hoarseness
- PMH
 - Unvaccinated (Mennonite)
 - No history of antibiotics or steroid course
 - No family history of malignancy or sudden pediatric death



Pertinent Physical Exam and Labs

ΡE

- General:
 - Alert, with obvious neck swelling visible from a distance; no apparent distress; audible noisy breathing with open mouth
- HEENT:
 - significant tonsillar hypertrophy (Mallampati 3-4)
- Neck:
 - 2 large, firm but mobile lymph nodes on left anterior cervical chain (3-4 cm in diameter), 3 additional smaller lymph nodes on the left (2-3 cm in diameter)
 - fullness in left supraclavicular area without palpable lymphadenopathy
 - \circ right side of the neck enlarged, with a 3-4 cm fixed nodal mass

Labs

- CBC
 - WBC 6.25
 - Hgb 10.8 (L)
 - Hct 31.4 (L)
 - RBC 3.89 (L)
- LDH 1,324 (H)
- Uric Acid 3.8



What Imaging Should We Order?



ACR Appropriateness Criteria

<u>Variant 2:</u>

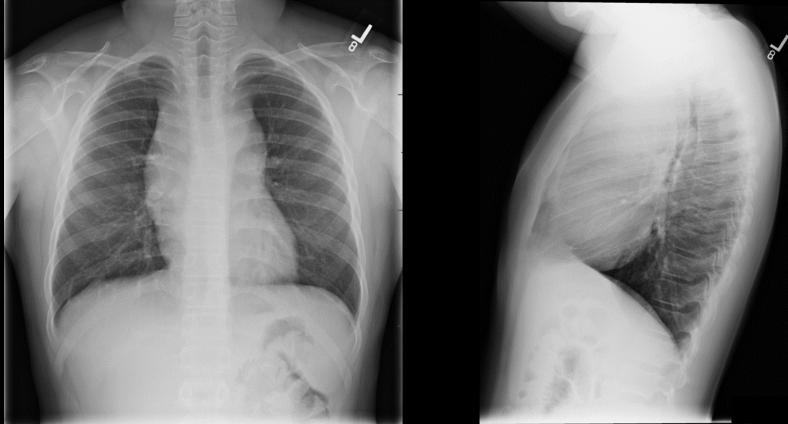
Child. 3 months of age and older. Immunocompetent. Community-acquired pneumonia that does not respond to initial outpatient treatment or requires hospital admission. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	•
US chest	May Be Appropriate	0
CT chest with IV contrast	Usually Not Appropriate	
CT chest without and with IV contrast	Usually Not Appropriate	
CT chest without IV contrast	Usually Not Appropriate	
MRI chest without and with IV contrast	Usually Not Appropriate	0
MRI chest without IV contrast	Usually Not Appropriate	0

This imaging modality was ordered by the admitting physician based on the patient presentation of ongoing cough and fatigue.

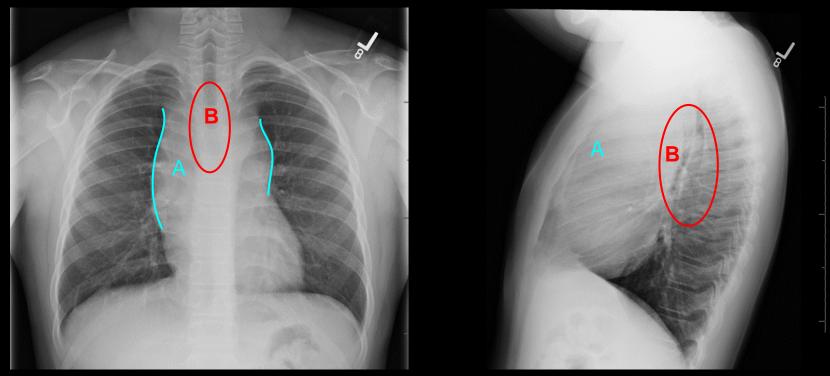


Findings (unlabeled)





Findings (labeled)



Opacified region of anterior mediastinum with widened transverse dimension for patient age; opacity is obscuring retrosternal clear space, concerning for a mass (A). There is mass effect on the trachea, which is flattened in anteroposterior dimension and narrowed and posteriorly displaced just above the level of carina in the lateral dimension (B).

Differential Diagnosis

- Lymphoma (Hodgkin, Non Hodgkin)
- Teratoma
- Thymic hyperplasia
- Thymic Cyst
- Thyroid (retrosternal extension)
- Malignant Germ Cell Tumor
- Thymoma
- Leukemia
- Lymphatic Malformation



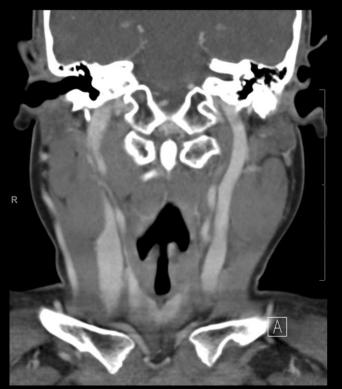
ACR Appropriateness Criteria

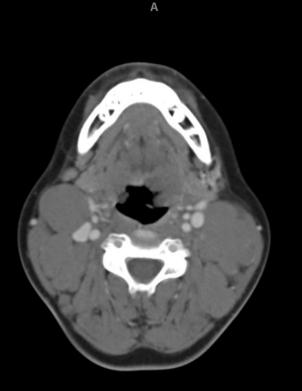
<u>Variant 4:</u> Child. Neck mass(es). Not parotid region or thyroid. Initial imaging.		
Procedure	Appropriateness Category	Relative Radiation Level
CT neck with IV contrast	Usually Appropriate	\$ \$ \$
MRI neck without and with IV contrast	Usually Appropriate	0
US neck	Usually Appropriate	0
MRI neck without IV contrast	Usually Appropriate	0
CT neck without IV contrast	May Be Appropriate (Disagreement)	• • •
MRA neck without and with IV contrast	Usually Not Appropriate	0
MRA neck without IV contrast	Usually Not Appropriate	0
CT neck without and with IV contrast	Usually Not Appropriate	* * * *
CTA neck with IV contrast	Usually Not Appropriate	* * *
Arteriography cervicocerebral	Usually Not Appropriate	* * * *
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	****
FDG-PET/MRI skull base to mid-thigh	Usually Not Appropriate	\$\$ \$\$ \$

This imaging modality along with chest CT with contrast were ordered by the admitting physician based on the cervical lymphadenopathy on clinical exam and chest radiography results.



Findings (unlabeled)

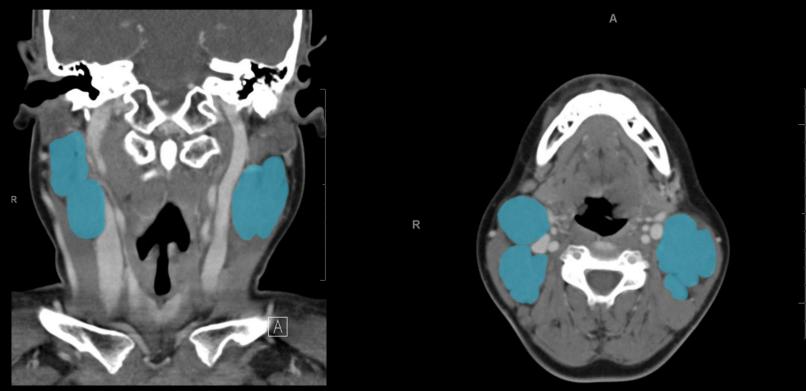






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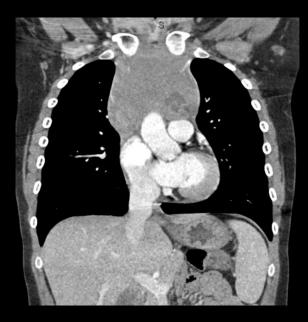
Findings (labeled)



Contrast enhanced CT of the neck showing enlarged tonsils and adenoids as well as multilevel lymphadenopathy including level IIa and IIb lymph nodes (blue).

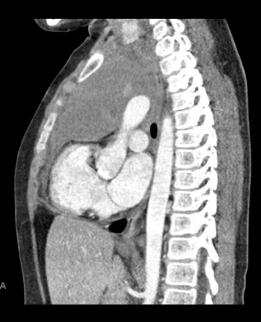


Findings (unlabeled)



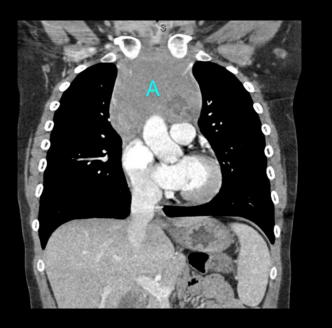


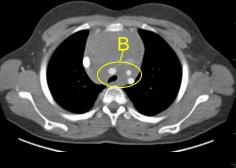


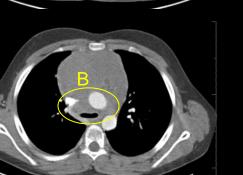


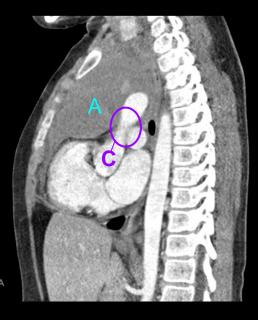


Findings (labeled)









Anterior mediastinal mass is identified which measures approximately $16.2 \times 5.6 \times 9.7 \text{ cm}$ (A). The mass encases the great vessels and compresses the trachea, which is flattened in anteroposterior dimension (B). The left brachiocephalic vein is narrowed in caliber as it crosses the mediastinal mass and enters the distorted superior vena cava area (C).



Pathology

Right upper cervical lymph node FNA and core biopsy

 Aspirate smears and cores are concordant with sheets of lymphoblasts.

Anterior mediastinal mass FNA

- Phenotype: Percentage of cells with abnormal phenotype: 97%
- Morphology: predominantly lymphoblasts and rare prolymphocytes

Bone marrow biopsy

• Less than 25% involvement



Final Diagnosis:

T-Cell Lymphoblastic Lymphoma



Anterior Mediastinal Masses in Children

The incidence of diseases presenting as an anterior mediastinal mass in children is different than in adults.

Differential - "The Four T's" :

- Terrible Lymphoma
- Teratoma
- Thyroid/Other
- Thymoma/Thymus

Lymphoma, usually Non Hodgkin (NHL), is the most common mediastinal mass in children and accounts for 50% of mediastinal masses in children compared to only 20% in adults.

Teratoma is the most common germ cell tumor in children but is less common in adults. Teratomas are generally found as mature benign teratomas, but a minority are immature malignant teratomas.

Thyroid disorders are rare in children. Lymphatic malformation/cystic lesions, metastases and infection are conditions that fall under "other."

Thymoma is very rare in children; however, normal thymus is prominent in early childhood and can be mistaken for a mass. Thymic hyperplasia is a benign condition more common in older children, often in response to an illness or medication (i.e., chemotherapy).

Imaging Workup for Mediastinal Masses in Children

Chest radiography may demonstrate the presence of a mediastinal mass, especially if it is large or displaces/compresses adjacent structures; however, it often does not allow for characterization of the lesion.

Cross sectional imaging with CT, MRI, or ultrasound allows for better:

Tissue characterization (solid, fatty, cystic, calcification, enhancement)

Assessment of size, location, spread

Effect on adjacent mediastinal structures.

While clinical history provides the background, information gained from imaging is key to diagnosis of mediastinal masses in the pediatric population.



Imaging Workup for Mediastinal Masses in Children

CT characteristics of common causes of anterior mediastinal masses in children:

- Lymphoma: Enhancing nodular soft tissue mass that may include areas of necrosis and cystic change. Often associated with regional lymphadenopathy and vascular or airway displacement or narrowing.
- Teratoma: Mixed cystic and solid lesions containing fat and calcification, displacing adjacent structures.
- Thyroid: Soft tissue mass extending from neck into anterior mediastinum. Thyroid tissue will demonstrate high attenuation (appear bright) without contrast due to normal physiologic concentration of iodine.
- Thymus: Bilobed, smooth soft tissue mass in the superior anterior mediastinum with mild homogeneous enhancement after contrast. The normal thymus will not displace adjacent structures. The normal thymus is prominent in children but involutes in adults.



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