AMSER Case of the Month November 2020

2 Year Old Male with Persistent Cough

Bathai Edwards, MS4 Penn State University College of Medicine

William W. Pryor, III MD Penn State University College of Medicine



James M. Brian, MD Penn State University College of Medicine



Patient Presentation

• HPI: 2-year-old with allergic rhinitis and eczema presents to clinic with chronic cough for the past 4 months. During this timeframe he had 2 suspected viral infections associated with runny nose and nasal congestion. He has not received relief from humidifier treatments, OTC cough medications or amoxicillin.

• ROS:

- Episodic posttussive emesis
- Nasal congestion
- Immunizations are up to date.



Patient Presentation

- Physical Exam:
 - Afebrile
 - General: Well-appearing in no acute distress
 - HEENT: Mild nasal congestion with clear discharge. Erythematous oropharynx with postnasal drip
 - Pulmonary: Coarse breath sounds in the upper lobes bilaterally

Pertinent Labs

• Nasal Respiratory Viral Panel positive for Influenza A and rhinovirus.



What Imaging Should We Order?



Select the applicable ACR Appropriateness Criteria

Variant 2:

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 referring physician







The trachea is midline with a right-sided upper mediastinal prominence (A). An airway indentation is seen along the right side of the trachea above the level of the carina (B).



Yellow arrow points to anterior bowing of the trachea







Next, the referring physician ordered an upper GI study due to the findings on chest radiography.



Barium esophagram showing posterior impression along the upper thoracic esophagus (B).



Next, CT angiography was ordered.









Axial CT angiography demonstrating double aortic arch surrounding the trachea esophagus (A). Sagittal view showing two large vessels anterior and posterior to the trachea causing compression (middle image and C), completing the vascular ring/double aortic arch.













Posterior view (A) and craniocaudal (B) view of CT angiography 3D reconstruction showing a dominant right-sided aortic arch (C) and smaller left-sided aortic arch (D) surrounding the airway and esophagus.



Final Diagnosis

Double Aortic Arch (Vascular Ring)



Vascular Rings

- Vascular rings are congenital aortic arch abnormalities that can cause tracheal and esophageal compression. They can occur due to embryological anomalies involving the 4th or 6th fetal aortic arches, the dorsal aortae, and/or the 7th intersegmental arteries based on the Rathke and Edwards models.
- Right aortic arch with an aberrant left subclavian artery is the most common type, followed by double aortic arch.
- Children may present at varying ages with noisy breathing, cough, or dysphagia due to compression of the trachea or esophagus. Additionally, patients may develop recurrent upper respiratory tract infections and dyspnea.



Imaging Workup

- Chest radiography may demonstrate a midline trachea with right aortic arch (in double aortic arch the right aortic arch is typically larger and more obvious) and tracheal bowing/narrowing, but further imaging is required.
- Barium esophagrams have been traditionally used and allow for visualization of posterior and right sided esophageal indentations consistent with a vascular ring.
- Cross-sectional imaging is often needed to accurately visualize the anatomy of the vascular ring and characterize its effect on adjacent structures. CT angiography is useful for surgical planning as it allows for complete assessment of the aortic arch and its branching vessel. 3-D rendered images derived from cross-sectional studies can provide even greater anatomical detail. MR angiography is also an option, without the need for ionizing radiation, although sedation is required and carries its own intendant risks.



Treatment

- Surgical intervention is indicated in **symptomatic** patients as the definitive treatment.
- Surgical correction provides symptomatic relief in most children.



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