RADY 403 Case Presentation: Vascular Ring

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May 2022
Focused patient history and workup

• 8 week old female with a pmh of Down Syndrome presented to a routine medical genetics office appointment with choking, spitting up, and regurgitation of food after most feeds

• Vitals:
  • Temp 36.3 °C (97.3 °F) (Temporal)
  • Ht 55 cm (21.65”)
  • Wt 4415 g (9 lb 11.7 oz)
  • HC 37 cm (14.57”)
  • BMI 14.59 kg/m²
  • BSA 0.26 m²

• Labs:
  • TSH 1.964
  • CBC: WBC 4.6, HGB 12.1, Plt 223, Absolute Lymphs 1.8
List of imaging studies

- Barium swallow 12/21/2021
- Echocardiogram 2/23/2022
- CTA Chest 5/10/2022
Barium Swallow
Barium Swallow

• Esophagram lateral projection shows **posterior esophageal compression** at the level of the second posterior rib
Barium Swallow
Barium Swallow
Barium Swallow

- Esophagram anterior projection demonstrates left-sided esophageal compression at the level of the 2nd posterior rib
Axial CTA Chest

- A-B: Aberrant right subclavian coursing inferiorly from the right axilla to the mediastinum. Note the posterior location relative to the esophagus in (B).
Axial CTA Chest

- C-D: Aberrant right subclavian continues to track inferiorly in the mediastinum from the right to the left side. Note the left sided location of three aortic branches in (C) and the arch in (D).
Axial CTA Chest

- E-F: The aberrant right subclavian joins the left sided descending aorta in the left mediastinum. Note the abnormal flattening, or pancaking, of the trachea.
Coronal CTA Chest

• A-C: Can see the more anterior right and left common carotid arteries in (A). Note the normal left subclavian in (B) and (C) with the origin of the aberrant right subclavian coming into view distally from the descending aorta in (C).
Coronal CTA Chest

- D-E: Moving posteriorly in the chest the aberrant right subclavian can be seen exiting the mediastinum in (D), and entering the axilla in (E)
Patient treatment or outcome

• Barium swallow demonstrated posterior esophageal compression on the left lateral view and left sided esophageal compression on the AP
  • Consistent with right aberrant subclavian

• Echocardiogram consistent with right aberrant subclavian, also incidentally found duplicated left sided SVC

• CTA of the chest performed for management planning confirmed diagnosis of left aortic arch with right aberrant subclavian and duplicated SVC

• The patient is currently pending treatment
Patient treatment or outcome

- General treatment for symptomatic right aberrant subclavian is surgical
  - The approach is individualized, but a common approach is division of the aberrant subclavian with anastomosis to the carotid (known as subclavian carotid transposition)
- Aneurysmal dilation is common at the origin of the aberrant artery and requires prophylactic surgery due to risk of aortic dissection
Vascular Rings General Information

- Vascular rings with compression of the trachea and esophagus comprise 1-3% of all congenital cardiac anomalies
- Divided into two categories
  - Complete rings: Double arch, Right arch/left ligamentum arteriosum
  - Partial rings: Pulmonary artery sling (aberrant left pulmonary artery), innominate artery compression, left arch with aberrant right subclavian

**NB:** The black structure is the trachea and the white structure is the esophagus.
Vascular Rings General Information

• Most common vascular rings
  • Right aortic arch and an aberrant left subclavian artery and left-sided ductus arteriosus = 30-65% of cases
  • Double aortic arch = 30-45% of cases
  • Innominate artery compression syndrome = 3-20% of cases
  • Aberrant left subclavian = 3-7% of cases
  • Pulmonary artery sling = <5% of cases
  • Aberrant right subclavian = <5% of cases

• NB: Aberrant right subclavian is the most common embryologic abnormality of the aortic arch, but is very uncommon with accompanying left sided aortic arch
Aberrant Right Subclavian With Left Aortic Arch

- Has an incidence as low as 0.07-0.2% in individuals with left sided aortic arch
- Symptomatic left arch with aberrant subclavian most commonly causes posterior esophageal compression WITHOUT tracheal compression
  - Can cause feeding difficulty called dysphagia lusoria
  - No issues with breathing

(Note! can be fooled thinking classic presentation of posterior esophageal course because 15% between trachea and esophagus and 5% anterior to the trachea rather than posterior)
Aberrant Right Subclavian Artery Associations

• Associated anatomic variations:
  • Truncus bicuspidus (common origin of carotid arteries) 19.2% of the time
  • Proximal aberrant subclavian aneurysm (Kommerell diverticulum) 14.9% of the time
  • Aneurysm of the distal aberrant subclavian 12.8% of the time
  • Right sided aortic arch 9.2% of the time
• Some studies suggest an incidence of up to 19-36% in children with down syndrome
  • Also suggested higher incidence in other trisomies such as 18
In pediatric patients, especially infants, differentiating oropharyngeal from retrosternal dysphagia is extremely challenging.

Patients with unexplained dysphagia should have a biphasic esophagram to look at structures from the oral cavity to the gastric cardia.

Lesions of the esophagus or gastric cardia can cause referred dysphagia sensation to the pharynx.
UNC Top Three

• Pediatric dysphagia should prompt consideration of a vascular ring
• Biphasic esophagram to look at structures from the oral cavity to the gastric cardia should be performed in unexplained dysphagia per the ACR appropriateness criteria
• Right aberrant subclavian is a specific prevalent ring that should be looked for in patients with Down Syndrome
Additional Finding – Duplicated Left SVC

- Duplicated SVC is seen coursing down the left side of the mediastinum rather than returning to the right heart via a normal left brachiocephalic vein.
Additional Finding – Duplicated Left SVC

- Moving from anterior to more posterior CT slices, the duplicated SVC drains into the coronary sinus, which is dilated as result.
References


• Amini, B., Molinari, A. Aberrant right subclavian artery. Reference article, Radiopaedia.org. (accessed on 19 May 2022) https://doi.org/10.53347/rID-831


References (cont)


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