Innominate Artery Repair: Treatment of Dysphagia Secondary to Vascular Compression

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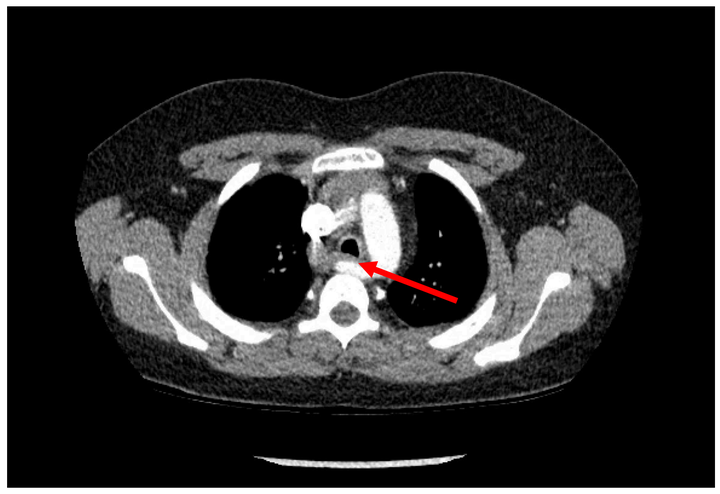
Background

- Innominate artery compression syndrome is a rare congenital anomaly due to an abnormal path of the artery leading to compression of the trachea and/or esophagus (complete or partial vascular ring)[1,5].
- Literature reporting repair focuses on symptomatic tracheal compression associated with dyspnea, cough, or recurrent infections[2].
- There is little focus on treatment of dysphagia via esophageal compression.
- Repair techniques focus on aortopexy or reimplantation of the innominate artery[7].
- There is often recurrence of disease and need for retreatment with revision of repair[3,6].
- In this case, esophageal compression dominated the symptoms, marked by dysphasia to solids.

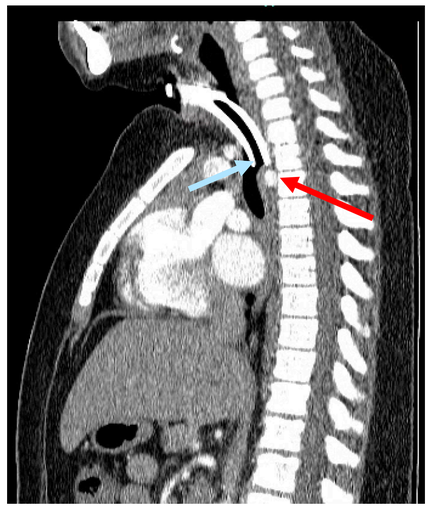
Case Description

- A 14-year old male with Trisomy 21 had severe chronic upper airway obstruction.
- After recovery from his adenotonsillectomy with direct laryngoscopy and bronchoscopy, polysomnography revealed persistent severe obstructive sleep apnea. This was attributed to obesity and laryngotracheomalacia, and he underwent recommended tracheostomy.
- He presented in out-patient follow up to the Pediatric Tracheostomy Clinic with reports of worsened severe dysphagia to solids. He was purported to "love salads, but was choking on the lettuce every time he ate one and had to vomit it up."
- His reported tracheomalacia finding combined with newly progressive severe dysphagia symptoms were further evaluated with Esophagram and Computed Tomography Angiography (CTA) of the chest.
- CTA revealed a left aortic arch with an aberrant right subclavian artery crossing posterior to the esophagus below the thoracic inlet.
- Esophagram revealed focal indentation on the in the posterior and left upper esophageal wall just below the level of the thoracic inlet, consistent with the vessel.
- Repair was performed by the pediatric cardiac surgeon via right thoracotomy, in which he translocated the aberrant right subclavian artery to right common carotid artery.
- The patient recovered well and had complete resolution of the dysphagia.

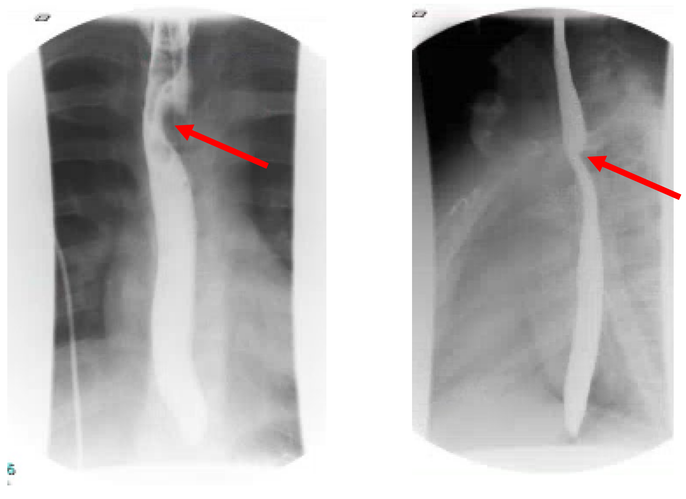
Imaging



↑ Figure 1: Computed Tomographic Angiography (CTA) of the chest reveals the aberrant innominate artery compressing the esophagus posteriorly (red arrow). Axial view using abdominal window.



← Figure 2: CTA of the chest revelas the aberrant innominate artery compressing the trachea and the esophagus posteriorly. Sagittal view using abdominal window. Note that the distal tracheostomy tube (light blue arrow) tip can be visualized to approach the path of the aberrant innominate artery (red arrow).



↑ Figure 3: Esophagram with posterior and lateral esophageal wall indentation near the thoracic inlet (red arrow). Coronal view (left) and Sagittal view(right).

Discussion

- Most congenital partial and complete vascular rings are diagnosed in infancy. Some remain asymptomatic for years.
- In infancy, the primary concern with vascular rings is respiratory symptoms with / without decompensation, such as cough, wheezing, respiratory failure, recurrent infections. The feeding symptoms presenting on the infant's liquid diet are usually secondary to respiratory symptoms[4].
- Isolated feeding symptoms of dysphagia are often not the focus of follow up symptomatic surveillance, nor are they often considered in isolation as the indication for surgical repair of innominate artery compression syndrome.
- Therefore, this dysphagia secondary to compression is often underdiagnosed and undertreated in the pediatric patient population.
- Surgical treatment is not novel, and the risks and benefits of repair must be discussed based upon the patient's particular circumstances.
- As in the case presented, the patient's quality of life can be improved. For patients who are potentially liquid-diet dependent due to symptoms, meeting nutritional needs with potentially unnecessary dietary restrictions creates a social stigma, a logistical challenge, and a safety concern.
- This case displays the utility of surgical correction of the vascular ring anomaly for the treatment of dysphagia to solids.
- Future research should focus on dysphagia as an indication for surgical repair, particularly elucidating which surgical treatment approach has the best results with the least risk. Other research should focus on outcomes that include quality of life improvements for patients with dysphagia that resolves after surgery.

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