ABERRANT RIGHT SUBCLAVIAN ARTERY PRESENTING AS UPPER AIRWAY OBSTRUCTION AND DYSPHAGIA LUSORIA

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INTRODUCTION

- An aberrant right subclavian artery is an exceedingly rare condition that falls under the umbrella of congenital vascular rings
- Involvement of the tracheobronchial tree and esophagus dictates symptomatology
- Medical literature is limited, however there appears to be a bimodal distribution in the age of presentation

CASE PRESENTATION

- A 42-year old female with history of cerebral palsy and asthma presented to the emergency room with 10 days of wheezing despite albuterol treatment, followed by 1 day of postprandial coughing
- CT chest angiography showed significant tracheal luminal collapse at thoracic inlet and aberrant subclavian artery
- On examination, she was in acute respiratory distress, tripoding with audible stridor and evident sialorrhea, in addition to diffuse end-expiratory wheezes
- She was transferred to ICU and intubated. Several attempts to extubate her were futile and she underwent placement of tracheostomy and PEG tube
- Following tracheostomy, the patient was again extubated and remained stable on 6 LPM oxygen via trach mask
- Cardiothoracic surgery and interventional cardiology were consulted and performed right carotid to right subclavian artery bypass graft, followed by plugging of the aberrant subclavian artery
- Further review of the patient's CT scan showed a cervical spine osteophyte causing countercurrent compression of the aberrant Subclavian artery, and subsequently near-total occlusion of the esophagus and significant compression of the tracheal lumen
- The patient was discharged home with close follow up

Figure 1 and 2.
CT Angiogram of the pulmonary vasculature. In figure 1, the origin of the aberrant R subclavian artery off of the aorta is seen, coursing posterior to the esophagus and trachea. In figure 2, a cervical spine osteophyte is seen causing anterior mass effect on the aberrant R subclavian artery and subsequent near-total occlusion of the esophagus and membranous trachea

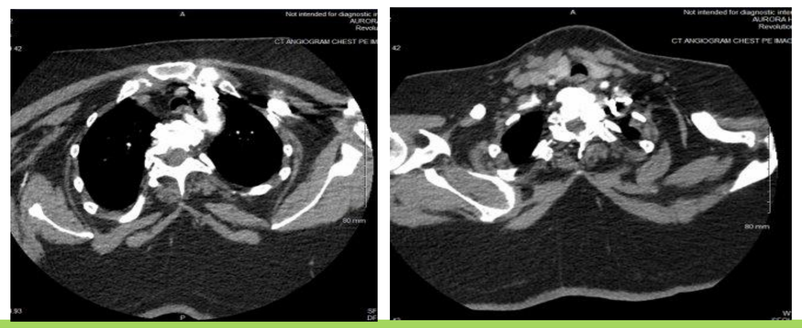


Figure 3.
Outline of interventions completed by interventional cardiology and cardiothoracic surgery to treat the anatomical defect.

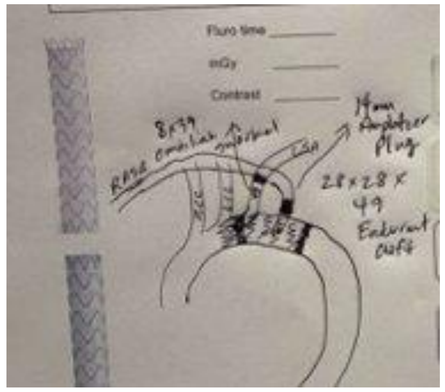


Figure 4a and 4b.
Outline of normal subclavian anatomy vs aberrant subclavian anatomy

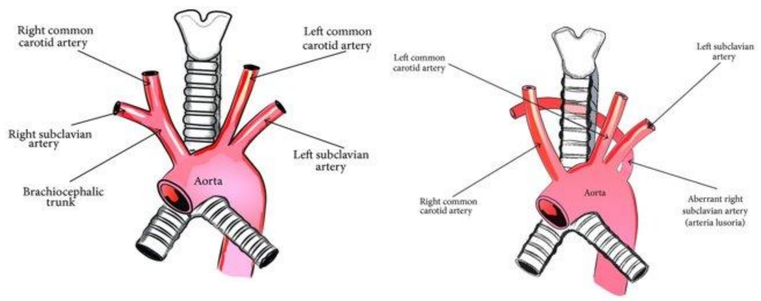
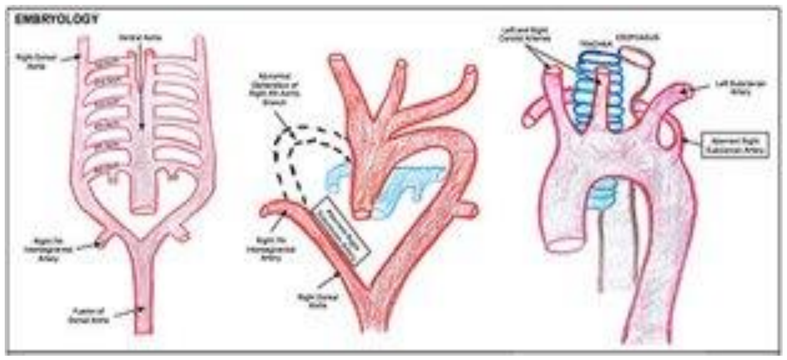


Figure 5.
Embryology: Beginning at the fourth week of embryogenesis, the aortic arch develops from six symmetrical paired aortic arch vessels and the paired dorsal aortae. During the next few weeks of embryogenesis, remodeling and rearrangement of these structures result in the formation of the normal left aortic arch.



DISCUSSION

- This case illustrates a congenital anatomical abnormality manifesting in a hyper-acute presentation. Diagnoses include:
  - Acute on chronic upper airway obstruction with excessive dynamic airway collapse
  - Dysphagia lusoria, a term used when a congenital vascular anomaly of the aortic arch and major branches is a cause of dysphagia
- The most commonly reported symptoms with congenital vascular rings include:
  - Dysphagia (71.2%)
  - Dyspnea (18.7%)
  - Retrosternal pain (17.0%)
  - Cough (7.6%)
  - Weight loss over 10 kg over a 6-month period (5.9%)
- Symptoms are present in only 7-10% of cases, and prevalence is higher in Down's, DiGeorge, and Edwards' syndromes
- Symptoms occur at the two extremes of life:
  - In children, tracheal obstruction or dysphagia can occur. The increased frequency of pulmonary infections seen in infants is thought to be due to the absence of tracheal rigidity
  - In infants, the trachea is compressible; therefore, the typical signs and symptoms compression by arteria lusoria are respiratory, such as wheezing, stridor, recurrent pneumonia, and cyanosis
  - In adults, the trachea is more rigid, and so, respiratory symptoms are rare
- Treatment with open surgery or endovascular stent-grafting is indicated for symptomatic relief and for prevention of complications due to aneurysmal dilatation
- This patient was placed on dual antiplatelet therapy with aspirin and clopidogrel for a minimum of 1 year

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