Dysphagia, Dyspnea, and Diverticulum: A Right Sided Aortic Arch with Aberrant Left Subclavian Artery

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Introduction:
- A right sided aortic arch is a rare congenital defect that can induce dyspnea with any resulting tracheal compression. The Edwards classification describes the anatomy of an aberrant left subclavian artery, which can arise from Kommerell’s diverticulum, a remnant of the left dorsal aortic root. When this diverticulum is aneurysmal, it can also induce dysphagia due to posterior esophageal compression.

Case History:
- A 27 year old female with a history of asthma requiring 2 previous intubations within the last year, vocal cord dysfunction, anxiety and bipolar depression presented to the emergency department with acute onset dyspnea.
- Patient denies ever using tobacco or vaping.
- She presumes it is asthma exacerbation, triggered by cigarette smoke from a bystander as is consistent with her history.
- On further questioning, she also endorsed experiencing occasional difficulty swallowing.
- She denies any stressors and admits to having panic attacks at home but notes this feels different.

Examination:
- Vitals on arrival to the ED: BP 114/78, HR 124, RR 26, SPO2 90% room air.
- On lung auscultation, she was noted to have rhonchi, wheezing and stridor.
- Cardiac work up revealed sinus tachycardia.
- Benign blood work and CXR without consolidation, a widened mediastinum with a right sided aortic knob.
- Initial bloodwork revealed respiratory alkalosis consistent with her tachypnea.
- In the ED, she had markedly increased work of breathing, tachypneic with rates in the 50s requiring BiPAP, and was admitted to the ICU.
- She was continued on her home montelukast and inhaled corticosteroid, and was admitted for systemic corticosteroids as well as nebulizer treatments.
- Though never developing significant hypoxemia, she had relapsing periods of dyspnea and tachyypnea requiring continued intermittent daily BiPAP, which seemed refractory to continuous inhaled nebulizer treatments.

Diagnosis:
- A CT angiogram of the chest was obtained, revealing the presence of a right sided aortic arch with aberrant origin of the left subclavian artery.
- This was confirmed with high resolution CT chest with contrast.
- This raised concern for the presence of a tracheal ring, along with esophageal involvement from a suspected Kommerell’s Diverticulum.
- Due to her difficulty swallowing, a barium swallow was obtained, which revealed extrinsic compression of the proximal esophagus, consistent with esophageal compression caused by the Kommerell’s Diverticulum.
- Pulmonary function testing was ordered but aborted due to dyspnea.

Discussion:
- The right sided aortic arch is classified into 3 types based on order of arteries branching out from the aorta:
  - Type 1: Left innominate artery, right common carotid, and right subclavian artery (mirror of the left sided aortic arch)
  - Type 2: Aberrant left subclavian artery
  - Type 3: Isolated left subclavian artery. The left subclavian artery does not attach to the aorta and is connected to the pulmonary artery.
- This patient has a type 2 right sided aortic arch. In type 2, the right aortic arch and aberrant left subclavian artery always has a left sided ligamentum arteriosum, which forms a vascular ring. This is because ligamentum arteriosum connects to the left pulmonary artery at the root of the subclavian artery. This is what causes the tracheal and esophageal compression.
- This is what caused the significant respiratory distress and intermittent dysphagia in this patient.

Conclusion:
- Her history of persistent asthma requiring intubation is noteworthy given these findings, which may explain why her asthma exacerbations appear refractory to steroids and nebulizers.
- This validated the severity of the patient’s symptoms, which were thought to have a psychogenic component given her history of bipolar depression and anxiety. This in turn, improved the patient’s well being.
- If the patient does in fact suffer from reactive airway disease, it is likely exacerbated by her anatomical finding.
- The patient was discharged to home and underwent elective surgical correction of her aberrant anatomy.
- With the appropriate surgical correction, the patient may be able to avoid further endotracheal intubations or a prolonged length of hospital stay in the future.

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References: