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:

- Vital signs 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 tachypnea 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 findings.
- 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: