Case Presentation

The case presented is a 22-year-old United States Air Force Basic Military trainee who presented to the Emergency Department with atrial fibrillation with rapid ventricular rate. His rate was controlled with intravenous diltiazem and he spontaneously converted to normal sinus rhythm shortly after admission. He denies having similar palpitations in the past, but reports that he had developed chest discomfort and dyspnea during his military physical training activities. Prior to joining the military he indicates he was very sedentary. Review of systems were notable for only mild dysphagia with solid foods, although this was intermittent and he had never sought medical advice for this symptom before. His physical exam, electrocardiography (after spontaneous cardioversion), chest roentography, and transthoracic echocardiography were normal. Given exertional chest discomfort and dyspnea, he underwent a gated cardiac CT. This demonstrated normal coronary origins, but his left pulmonary artery took an anomalous course between the trachea and esophagus. This anomaly has been described in the literature as a pulmonary artery sling (PAS, Fig.) and was thought to be the etiology of his dysphagia and exertional dyspnea.

Figure. Axial contrast-enhanced CT image through the chest demonstrates an anomalous course of the left pulmonary artery between the esophagus posteriorly and the tracheobronchial tree anteriorly. There is associated narrowing of the main stem bronchi and compression of the esophagus.
Key imaging finding

Vascular Ring

Differential diagnoses

- Pulmonary Artery Sling (PAS)
- Double Aortic Arch
- Right Sided Aortic Arch with Aberrant Left Subclavian Artery

Discussion

Vascular rings are congenital anomalies that can present with a variety of pulmonary and gastroesophageal symptoms. Patients will usually present with symptoms of airway obstruction due to tracheal compression manifesting as stridor, recurrent infections, wheezing, and cough. Esophageal complaints are also common and include dysphagia and vomiting. CT or magnetic resonance angiography is the preferred method of diagnosing vascular rings.

Pulmonary Artery Sling.

PAS is a rare vascular anomaly first described by Glaevecke and Doehle in 1897.\(^1\) It has been proposed that a PAS forms by the connection of the left post-brachial plexus with the right pulmonary artery, resulting in the left pulmonary artery coursing posterior to tracheobronchial tree.\(^2\) While a PAS can occur in isolation, many patients have associated congenital heart disease (e.g. atrial septal defects, ventricular septal defects, or patent ductus arteriosus) or noncardiac anomalies (e.g. tracheal stenosis, tracheomalacia, or lung hypoplasia).\(^3,4\) Patients with PAS are typically symptomatic and present within the first year of life with signs and symptoms of respiratory obstruction,\(^3\) with two-thirds of the patients presenting in their first month of life.\(^2\) However, in some case series, up to 12% of patients with PAS are asymptomatic.\(^2\) Although extensive review of the literature shows no clear guidelines regarding specific indications for surgery, corrective surgery is the mainstay of treatment if life-limiting symptoms such as severe dyspnea, stridor, or dysphasia are present.\(^2,3\) Asymptomatic patients do not require surgery and have an excellent prognosis.\(^5\)

Double Aortic Arch.

With a double aortic arch, the ascending aorta arises normally but divides into two aortic arches as it leaves the pericardium. The arches join back together to form the descending aorta. This anomaly is due to the failure of remodeling during embryogenesis. Patients usually present with symptoms similar to a pulmonary artery sling.\(^6\) Esophagram will show bilateral impressions on the thoracic esophagus. CT angiography or MRI is preferred over esophagography or angiography prior to surgical management.\(^7\)

Right Sided Aortic Arch Arch with Aberrant Left Subclavian Artery.

This anomaly is due to the interruption of left aortic arch between the left common carotid artery and the left subclavian artery. The aorta descends to the right of midline and a posterior left subclavian artery arises from the diverticulum of Kommerell, which is a remnant of the left sided arch.\(^7\) This anomaly is associated with cardiac defects in about 5% - 10% of cases. Complications such as aneurysmal dilatation or dissection of the anomalous vessel may result in symptoms of dysphagia. Esophagram will show posterior vascular indentation and a right-sided arch impression. Cross-sectional imaging is required to confirm the anatomy and to visualize the diverticulum of Kommerell.\(^7\)
Diagnosis

Pulmonary Artery Sling

Summary

Pulmonary artery sling is a rare vascular anomaly that is usually symptomatic within the first year of life. Symptoms are usually respiratory and/or gastroesophageal and require surgical intervention. In patients with symptoms of dysphasia, barium esophagram can be used to exclude vascular ring. Asymptomatic presentation is less common and is usually found incidentally on CT or MRI. Asymptomatic patients typically have a good prognosis. Our patient had resolution of all his symptoms when he abstained from rigorous physical training exercises; he opted to separate from the military.

References


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