Adult Presentation of Anomalous Pulmonary Artery from the Descending Aorta: A Rare Cause of Exertional Chest Pain

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CASE PRESENTATION

A 20-year-old female with no previous past medical history, other than a recent evaluation for Ehlers-Danlos syndrome, presented to the emergency department for chest pain. She described exertional right-sided chest pain increasing over the previous two months. Vital signs were stable, and her physical exam was unrevealing. Electrocardiogram demonstrated poor R-wave progression in the precordial leads. Laboratory testing included an unremarkable complete blood count, comprehensive metabolic panel, and troponin. Chest radiograph was normal. Her history of possible Ehlers-Danlos syndrome prompted the emergency physician to complete a computed tomography angiogram (CTA) of the chest and abdomen for consideration of aortic dissection. The CTA revealed a pulmonary artery originating from the descending aorta above the celiac plexus, supplying the right lower lobe of the lung (Images 1 and 2). Outpatient follow-up with primary care, cardiology, and cardiothoracic surgery was ensured. Outpatient echocardiogram revealed mild tricuspid regurgitation. Definitive management of the anomalous vessel was accomplished by endovascular closure using a 10-millimeter (mm) x 7 mm Amplatzer vascular plug (Abbott Laboratories, Abbott Park, IL).

DISCUSSION

An anomalous origin of a pulmonary artery branch is a rare congenital abnormality that comprises 0.12% of all congenital heart defects. The majority of reported cases involve left pulmonary arteries originating from the ascending aorta. Less frequently, anomalous pulmonary arteries have been described arising from the descending aorta and occasionally the celiac artery. Our patient’s specific congenital abnormality is unusual because her pulmonary artery originates from the descending aorta, supplying her right lower lobe. Symptoms such as chest pain, hemoptysis, and exertional dyspnea may help identify patients with anomalous pulmonary vasculature. The high pressure of systemic blood in a low-pressure pulmonary system can result in right heart strain, pulmonary hypertension, and high-output cardiac failure. However, most cases are discovered in utero and are associated with other congenital anomalies. Cases that are
What do we already know about this clinical entity? Anomalous pulmonary artery is a rare congenital disease that typically presents early in life with congestive heart failure.

What is the major impact of the image(s)? These images are an example of a rare disease entity as a cause of chest pain and respiratory distress, which are common presenting symptoms in the emergency department.

How might this improve emergency medicine practice? Although rarely discovered in adulthood, recognition of anomalous pulmonary arterial supply can lead to expedited referral and definitive management.

The authors attest that their institution requires neither Institutional Review Board approval nor patient consent for publication of this case report. Documentation on file.

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REFERENCES