A Symphony of Anomalies: Isolated Pulmonary Artery Agenesis Meets Congenital Lung Hypoplasia

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Consent: As this is a case report, consent was obtained for the purpose of this paper.

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Key Clinical Message:

Isolated agenesis of pulmonary arteries with congenital lung hypoplasia is rare. It can be found in childhood or adulthood if asymptomatic. We present a patient with congenital right lung hyperplasia with an absent right pulmonary artery.

Case Presentation:

A 24-year-old Female with a past medical history of intermittent asthma, right lung hypoplasia, and dextrocardia presented to the Emergency Department for evaluation of shortness of breath. Upon evaluation via imaging, she was noted on CT Angiogram (Figures A,B &C) to have congenital hypoplasia of the right lung, dextrocardia, and absent right pulmonary artery and its branches.

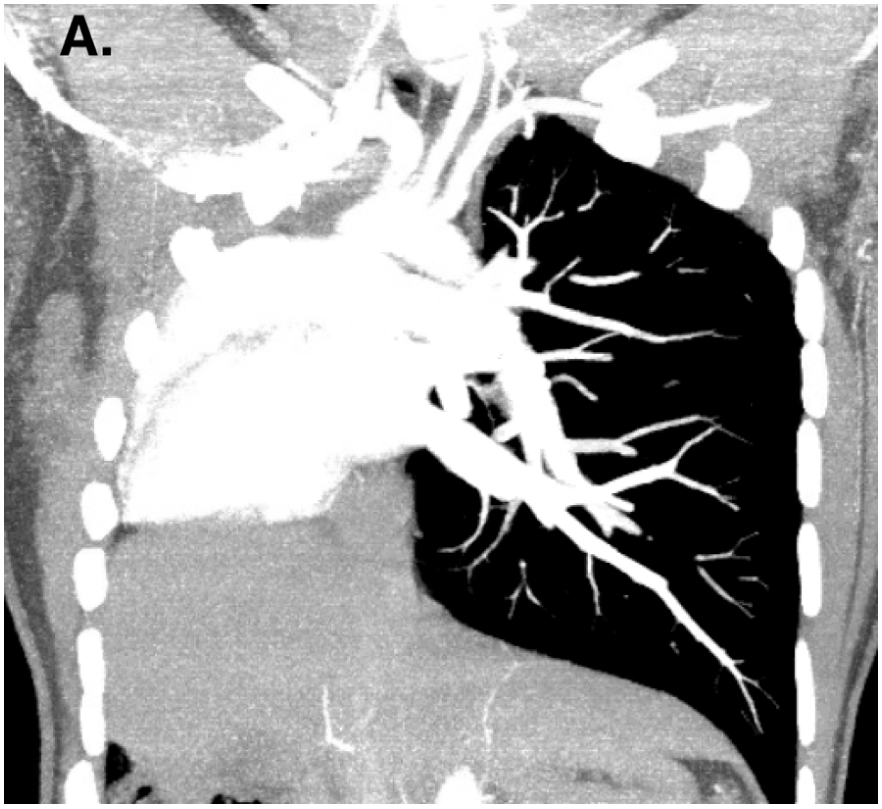

Figure A : Coronal View of Computer Tomography Angiography of the Chest demonstrating Absent Pulmonary Artery, Pulmonary Hypoplasia and Dextrocardia.

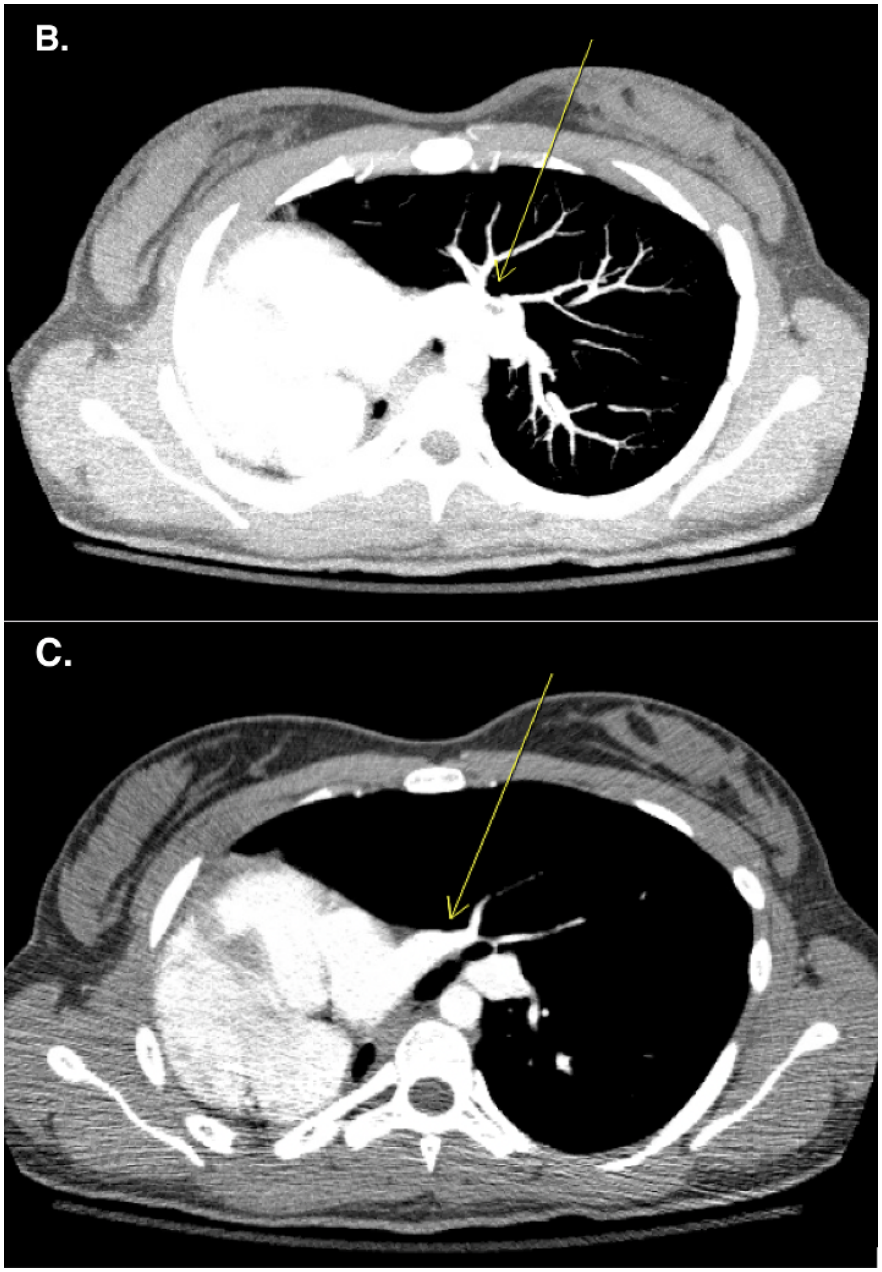

Figure B&C: Axial Views of Computer Tomography Angiography of the Chest demonstrating Absent Pulmonary Artery, Pulmonary Hypoplasia and Dextrocardia.

Discussion:

Congenital, isolated agenesis of pulmonary arteries (UAPA) is a developmental malformation that shows a unilateral absence of the pulmonary artery without the presence of a congenital heart disease [1]. This often leads to pulmonary hypoplasia of the ipsilateral lung [1]. UAPA is often classified as congenital or acquired [1]. Congenital is defined as a developmental defect or malformation during embryogenesis [1,2]. Acquired due to chronic subclinical infection, typically secondary to tuberculosis, that involves the hilum of the lung [1]. UAPA has a bimodal distribution in regard to age of presentation [1]. Those who are present in

childhood are often symptomatic as a result of congestive heart failure or pulmonary hypertension [1]. Those who present as an adult are often asymptomatic, typically due to collateral vasculature to the affected lung [1,2]. While there are many modalities that can be used for diagnosis, CT or MR pulmonary angiography is considered the gold standard [1].

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