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Case report

Unilateral pulmonary artery atresia in an adult: A case report

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ABSTRACT

Intro: Unilateral pulmonary artery atresia (UPAA), while encountered frequently in the congenital cardiac anomaly cohort, is occasionally diagnosed in adulthood after typical symptoms of hemoptysis, pulmonary infection, or as an incidental finding on contrast CT scan. Due to its rarity, a brief discussion of UPAA and its treatment is warranted.

Case report: A 35 year old male presented with three days of hemoptysis. After diagnosis of right UPAA, he underwent angioembolization of 6 large systemic collaterals supplying his right lung, followed by right pneumonectomy. He was discharged on post-operative day 3, and at follow up 6 weeks later was doing well with minimal residual incisional pain and excellent pulmonary reserve.

Conclusions: UPAA presents classically with hemoptysis, but also with pneumonia, pulmonary hypertension, or incidentally. Management includes selective collateral embolization, pneumonectomy, or medical management directed towards decreasing pulmonary hypertension in patients unable to tolerate pneumonectomy due to comorbidities. Pneumonectomy in these patients is characterized by dense and hypervascular adhesions, with large volume blood loss expected during adhesiolysis, which can be decreased with pre-operative embolization. Outcomes are typically excellent in otherwise healthy patients.

1. Introduction

Unilateral pulmonary artery atresia (UPAA) is a defect thought to be caused by failure of the embryonic sixth aortic arch to fuse with the pulmonary trunk during development [1]. While often encountered in the pediatric cardiac anomaly cohort, it is occasionally diagnosed in adulthood as an incidental finding or, more commonly, during a workup for hemoptysis. Though this diagnosis is exceedingly rare to see in clinical practice, knowledge of its clinical course and treatment are vital for safe and efficient care of this patient population. Consequently, we describe a patient with unilateral pulmonary artery atresia, summarize the current literature, and provide our practice's recommendations for treatment.

2. Case Report

Our patient was a 35 year old male who was admitted to our facility for three days of mild to moderate hemoptysis. The patient informed us that he had been told that he had an “undeveloped right lung,” and that he had suffered pneumonia and bronchitis several times in the past, but was otherwise healthy. A computed tomography scan of the chest with intravenous contrast revealed an absent right sided pulmonary artery (Fig. 1), with both smaller parenchymal volumes and multiple systemic collaterals on the right.

Prior to performing a right pneumonectomy, our interventional radiology team performed embolizations (Fig. 2) of four right intercostal arteries and two bronchial arteries that appeared to be supplying the majority of blood supply to the right lung.

The patient underwent a right pneumonectomy via a posterolateral thoracotomy with thoracoscopic assistance to aid in dissection of abundant dense adhesions and large collateral blood vessels. Approximately 1500 cc's of blood was lost during the procedure, however the patient tolerated the procedure well with no blood product transfusions required, and was discharged post operative day three in excellent condition. At follow up 2 weeks and 6 weeks later, he was recovering well with no oxygen requirements and tolerating his regular activities with no limitations.

3. Discussion

A total of 350 total cases have been reported in the literature [2–5], with 65 reported since 1990. Adult patients with unilateral pulmonary atresia typically present with hemoptysis or exertional dyspnea, but can also have recurrent respiratory infections, chest pain, or even a

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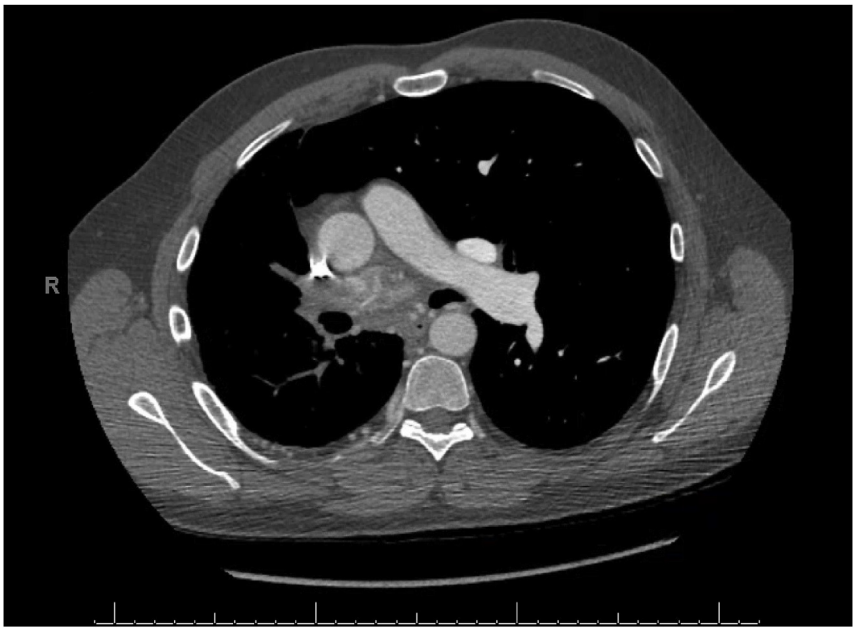

Fig. 1. Computed tomography at the level of expected pulmonary artery bifurcation showing absence of the right pulmonary artery.

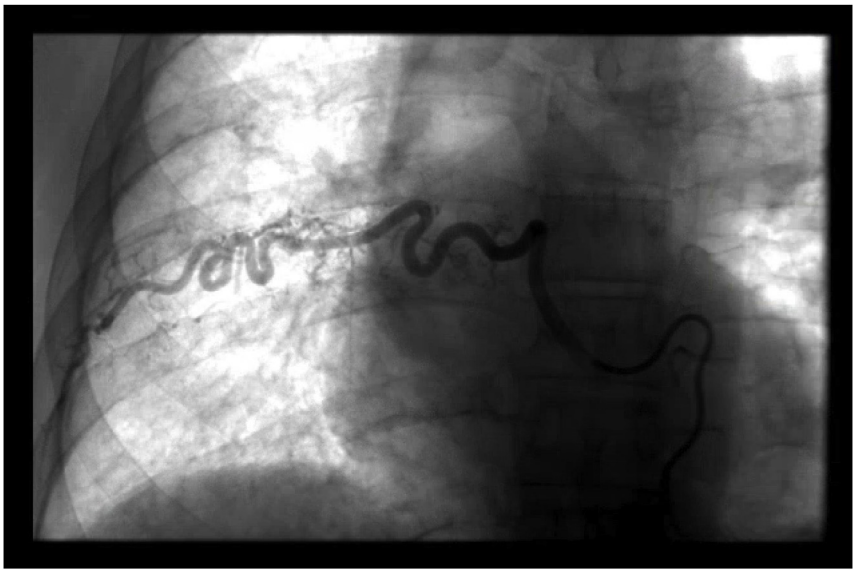

Fig. 2. Dilated bronchial artery collateral.

propensity for high altitude pulmonary edema. The lung parenchyma on the affected side can have bronchiectasis (particularly in patients with recurrent infections), interstitial changes, or multiple bullae [6].

In keeping with an embryonic developmental abnormality as the root cause, just under half of patients with an absent left pulmonary artery will have a right sided aortic arch, while those with an absent right pulmonary artery typically have a normal aortic arch. Collateral pulmonary flow develops from many sources in the thorax, including bronchial arteries (~70%), phrenic arteries (47%), the internal thoracic artery (44%), intercostal arteries (44%), the subclavian or axillary artery (34%), directly from the aorta (13%), and esophageal branches (9%) [6].

Therapies are directed first towards stabilizing the patient, with securing of the airway in unstable patients, and immediate management of any severe bleeding that could compromise ventilation. For the few patients that present in extremis, prompt intubation with bronchial blocking of the bleeding lung can be life saving. Angioembolization of active bleeding is then undertaken. Symptoms of bleeding and exertional dyspnea can be managed by selective angioembolization of large collaterals and/or medical management of pulmonary hypertension with beta-blockers, calcium channel blockers, or phosphodiesterase inhibitors in patients unable to tolerate surgical therapy. Pneumonectomy offers the best long term therapy in patients able to tolerate the procedure. While this may seem somewhat extreme, patients with a unilaterally absent pulmonary artery are functioning with little gas exchange occurring in the affected lung, so removal of the abnormal lung usually does not significantly affect their functional status. Consequently, pulmonary function tests and oxygen exchange evaluations should be interpreted cautiously, recognizing that the usual drop by 50% in gas exchange and FEV1 is not expected post-pneumonectomy. In asymptomatic patients, close surveillance is also an appropriate approach, provided the patient is reasonably able to understand the potential for future problems and has ready access to adequate medical care.

Performing a pneumonectomy in these patients is not straightforward. Due to the decreased perfusion in the affected lung, angioneogenesis results in a plethora of systemic supply via adhesions throughout the pleura. A large amount of blood loss is anticipated, with most of the blood loss occurring prior to encountering the hilar vessels and main bronchus. Video assistance in managing the apex and posterior lung is helpful, as is pre-operative embolization of collaterals. Typed and crossed blood is appropriate pre-operative preparation, but in healthy patients like ours, moderate volume blood loss is well tolerated provided adequate resuscitation is provided, including fluids and cell salvage. Post-op management is undertaken as usual for a pneumonectomy, with no suction applied to the chest tubes post-operatively, and early removal of drains to prevent mediastinal shift.

4. Conclusions

UPAA presents classically with hemoptysis, but also with pneumonia, exertional dyspnea, or incidentally. Management includes selective collateral embolization, pneumonectomy, and medical management directed towards decreasing pulmonary hypertension in patients unable to tolerate pneumonectomy due to comorbidities. Pneumonectomy in these patients is characterized by dense and hypervascular adhesions, with large volume blood loss expected during adhesiolysis, which can be decreased with pre-operative embolization. Outcomes are typically excellent in otherwise healthy patient.

Conflicts of interest

The authors have no competing interests to declare.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2018.11.012.

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