Case report

Unilateral pulmonary vein atresia: A rare case of hemoptysis

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A B S T R A C T

We present a rare case of hemoptysis secondary to isolated unilateral pulmonary vein atresia. Isolated pulmonary vein atresia is a rare condition in which patients typically acquire a diagnosis in infancy and early childhood [Mataciunas et al.; Pourmoghadam et al.]. Our patient presented during puberty with several previous episodes of hemoptysis prior to her admission and diagnosis. The initial diagnosis was suspected in our patient from chest computerized tomography (CT), and confirmed with cardiac catheterization and pulmonary angiography. Treatment aim is to preserve lung function and minimize irreversible pulmonary remodeling [Pourmoghadam et al.; Harrison et al.]. Conservative monitoring can be considered with milder or asymptomatic cases, while others may require preoperative collateral artery banding, surgical anastomosis between the pulmonary vein (PV) & left atrium (LA) and even pneumonectomy [Pourmoghadam et al.].

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1. Introduction

The presence of hemoptysis in the pediatric population is a rather uncommon complaint that can range on a spectrum of severity [4–6]. Initial workup should include complete blood count and chest radiography [4–6]. A chest CT is used to further delineate chest radiographic findings [1,7].

Isolated unilateral pulmonary vein atresia is a rare condition [3,8]. Patients generally present with recurrent pneumonia and hemoptysis [1]. Typically diagnosis occurs during infancy and early childhood [1]. This anomaly occurs due to a failure of incorporation of the common pulmonary vein into the left atrium, with 50% associated with congenital cardiac anomalies [2,3]. Due to risk of pulmonary hemorrhage and pulmonary hypertension, there is significant morbidity and mortality associated with this anomaly [2,3].

2. Case description

A 12-year-old African American female with a medical history significant for asthma and history of recurrent pneumonia presented to the emergency department with a complaint of hemoptysis, described as “coughing up a handful of bright red blood.” This episode of hemoptysis occurred after riding her bike and was associated with chest discomfort. The past medical history was significant for two previous episodes of scant hemoptysis, recurrent pneumonia and asthma. Physical exam was only remarkable for diminished breath sounds over the right lower lung field. The patient had age appropriate vital signs and was maintaining adequate oxygenation on room air.

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In the emergency department (ED), a complete blood count and coagulation panel was negative for any abnormalities. Initially, a chest radiograph was reported as normal. Therefore, a computed tomography (CT) scan of the chest was performed. The images on the CT were suggestive of hypoplastic right lung, as well as hypoplastic right pulmonary artery (RPA) [Fig. 1]. Therefore, the patient was admitted to the hospital for further evaluation and diagnostic studies.

In the hospital, a bronchoscopy and echocardiogram (ECHO) were performed, with the ECHO concerning for increased right ventricular systolic pressure. Bronchoscopy revealed diffuse erythema of the mucosa at the entrance to the bronchus intermedius of the right upper lobe [8], and too numerous to count hemosiderin laden macrophages. The decision was made to further evaluate the patient with cardiac catheterization, which showed the right pulmonary veins were very small and came to a confluence near the left atrium, but did not enter the left atrium [Fig. 2]. Multiple collaterals were coiled.

In our patient’s case, the anatomy of her anomaly supported anastomosis. The patient underwent a repair procedure three months later. The procedure consisted of repair of the right pulmonary vein atresia with excision of a membranous septum and a...
pericardial patch angioplasty of the right pulmonary venous confluence. The patient no longer requires asthma medications and has been weaned off anti-hypertensive medication.

3. Discussion

The presence of unilateral, isolated pulmonary vein atresia is rare [3,7]. Since patients present in early infancy or childhood, the age of our patient makes this case even more unusual [1,2]. This condition should be considered in children with hemoptysis, recurrent pneumonia and imaging indicating ipsilateral hypoplastic lung [9].

Diagnosis can be confirmed with cardiac catheterization if index of suspicion is elevated [9,10]. Treatment aim is to prevent co-morbid conditions and preserve lung function [2,3,9]. The approaches include conservative monitoring, surgical anastomosis or pneumonectomy with preoperative collateral vessel banding [2,9]. Aggressive treatment is dependent upon the severity [2,9]. Due to the morbidity and mortality of this condition, it is crucial to identify these patients before pulmonary hypertension or massive pulmonary hemorrhage ensues [2].

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