UNILATERAL PULMONARY VEIN ATRESIA - A CASE REPORT

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Abstract:
Unilateral pulmonary vein atresia without associated congenital cardiac anomalies is a rare condition. Patients with congenital unilateral pulmonary vein atresia are usually symptomatic and present from infancy or childhood with recurrent pulmonary infections or hemoptysis. But a subclinical course of the disease is rare and only few cases have been reported.

Keyword: pulmonary vein atresia, congenital cardiac anomaly

A 14 year old boy was admitted in the cardiology ward with gradually increasing shortness of breath for 2 months. One year back he was admitted in the medical ward for evaluation of hemoptysis. At that time he was suspected to have pulmonary tuberculosis but found to be negative and discharged. On examination, he was tachypneic at rest, anaemic, not cyanosed, with no clubbing or pedal edema. Vitals were stable; systemic examination revealed loud P2, rhonchi and crackles over both sides of chest. Routine investigations were normal except Hemoglobin of 9.2 g%. His chest Xray showed a dilated main pulmonary artery with peripheral pruning with right ventricular enlargement. ECG showed a Right axis deviation, Right atrial enlargement & Right ventricular hypertrophy with strain pattern.

Echocardiography showed a turbulence at the site of right upper pulmonary vein joining the Left Atrium; Left sided pulmonary veins could not be visualised; Tricuspid regurgitation was mild with a pressure gradient of 86 mmHg; Pulmonary Regurgitation mild with peak gradient of 40mm Hg.

Figure 1: Chest X ray
Figure 2: Echo PLAX view
Figure 3: Echo A4C view

Cathstudy showed a dilated Main Pulmonary Artery (MPA), Right Pulmonary Artery (RPA) and proximal Left Pulmonary Artery (LPA). Distal LPA was hypoplastic. Left pulmonary veins were not visualised; right pulmonary veins were seen draining into the left atrium. Both septae were intact. Pressure data revealed elevated systolic pressure of pulmonary artery to about 75% of the systemic pressure and elevated diastolic pressure. Pulmonary Capillary Wedge Pressure (PCWP) was elevated with a gradient between PCWP and Left atrial pressure; very mild desaturation in Left atrium suggested mild Left to Right shunt across Persistent Foramen Ovale.

CT Pulmonary angiogram showed a dilated MPA, RPA and LPA; LPA smaller than the RPA. Left atrium received right superior and inferior pulmonary veins; both left sided veins were not visualised and their insertion site in left atrium showed a bulge. Emphysematous changes of lungs seen on the left lung more than the right lung.
Discussion:
Congenital atresia of individual pulmonary veins without associated structural abnormalities of the heart is a rare anomaly. To date, there have been reports of 16 cases in English.\textsuperscript{1-3} Patients usually present with recurrent pulmonary infections or haemoptysis. Congenital atresia of the pulmonary veins is thought to be caused by defective incorporation of the common pulmonary vein into the left atrium leading to obstruction in some or all of its branches. Edwards has postulated that interruption of embryogenesis at different stages may be responsible for the variety of lesions seen\textsuperscript{4}. In rare instances the venous obstruction may be acquired—for example secondary to venoocclusive disease or mediastinitis. Shrivastava et al described the unusual association of unilateral pulmonary venous atresia with pulmonary veno-occlusive disease in the contralateral lung in a 7 year old patient who died\textsuperscript{3}. There was no associated congenital heart defect in this patient. Beerman et al reported a patient in whom pulmonary venous obstruction did not develop until some years after ductal ligation, after normal pulmonary venous drainage had been seen before operation. Beerman et al did not suggest a possible cause for the obstruction\textsuperscript{5}.

The typical presenting features were noted in our patients. In our patient an episode of haemoptysis and recurrent chest infections with wheezing were witnessed. Smaller hilar vessels on the affected side as seen in our patient. Echocardiography, catheterization and CT angiogram supported the clinical impression of pulmonary vein atresia. Many patients with pulmonary vein atresia have severe pulmonary hypertension as in our patient. In our patient it was possible to obtain a satisfactory wedge pressure on the left side and was found to be increased. We assume that the increased blood flow through the bronchial arteries which normally carry less than 5% of the cardiac output and communicate with the pulmonary branches via precapillary anastomoses was responsible for the raised pulmonary wedge pressure, that was found on the affected side. Pulmonary angiography confirmed the diagnosis in our case. Selective injections into the pulmonary arteries showed slow passage of contrast into the affected lung with failure to demonstrate pulmonary veins draining into left atrium on that side. Without surgical treatment mortality in this condition approaches 50%. Surgical treatment mainly comprises pneumonectomy, because a direct approach on the pulmonary veins is usually not feasible.

Conclusion:
We describe this patient with congenital atresia of the pulmonary veins and confirm the value of pulmonary angiography in clarifying the diagnosis of this rare condition. The diagnosis should be considered in any child with recurrent unilateral pneumonia, especially if it is associated with haemoptysis and radiographic evidence of reduced lung volume on the affected side.
References:

