Anomalous origin of pulmonary artery from aorta complicated by pulmonary thrombosis: diagnosed using echocardiography

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A 35-year-old male was admitted due to increasing dyspnea and chest pain. Physical examination found cyanosis, clubbed fingers, and systolic murmur at the left sternal border. Laboratory tests showed polycythemia (hemoglobin 221 g/L) and hypoxemia (PO2 50.8 mmHg). Echocardiography revealed that there was discontinuity of the pulmonary artery branches with the left pulmonary artery (LPA) originating from the main pulmonary artery and the right pulmonary artery (RPA) from the ascending aorta. RPA was severely dilated with a large echogenic mass suggestive of a large thrombus (Panels A and B). A large patent ductus arteriosus (PDA, diameter 11 mm) was present between the LPA and descending aorta (Panel C). Severe pulmonary hypertension was present in both pulmonary artery branches as suggested by a high velocity tricuspid regurgitation jet (around 6 m/s) and right-to-left shunting through the PDA (Panel D). CT angiography confirmed the echocardiographic findings, and confirmed the presence of a large thrombus in the RPA together with prominent arteriopulmonary collaterals (Panels E and F).

Anomalous origin of one pulmonary artery from aorta (AOPA) is a rare congenital anomaly, often associated with other heart malformations. Patients usually develop severe pulmonary hypertension in early age, with a mortality of 80% at 1-year-old. AOPA in adult is extremely rare. It has never been reported to be complicated by pulmonary thrombosis. The mechanism for the pulmonary thrombosis in this case might be polycythemia and slow flow velocity secondary to significant pulmonary hypertension and dilatation. Early diagnosis usually provides chances of successful surgical correction of AOPA. In adult patients, irreversible obstructive pulmonary hypertension is the main contraindication for surgical correction, and combined heart-lung transplantation may be the only solution for these patients.