Medical Image of the Week: Eisenmenger Syndrome and Hemoptysis

A 26 year-old female with Eisenmenger syndrome presented with hemoptysis. An echocardiogram showed an enlarged right ventricle and two large mid-muscular ventricular septal defects (VSD) with right to left shunting (Figures 1 and 2).

Figure 1. Apical four-chamber view of the heart as seen on a transthoracic echocardiogram demonstrating an enlarged right ventricle (RV) and two large mid-muscular ventricular septal defects (*). RA - right atrium, LA - left atrium, LV - left ventricle.

Figure 2. Apical four-chamber view on a transthoracic echocardiogram. Color Doppler jets (blue color) demonstrate right-to-left shunt through the two mid-muscular ventricular septal defects seen in Figure 1.
A contrast enhanced CT of the chest showed an enlarged pulmonary artery, no evidence of pulmonary embolism and the VSDs (Figure 3 and 4).

Figure 3. Contrast enhanced CT of chest demonstrating markedly enlarged main pulmonary artery (arrow), approximately twice the size of the ascending aorta (straight arrow).

Figure 4. Contrast enhanced CT of chest showing ventricular septal defects (arrows).
Eisenmenger syndrome is a condition in which increased pulmonary blood flow secondary to a left to right intracardiac shunt leads to irreversible pulmonary vascular obstructive disease. The resultant high pulmonary vascular resistance causes reversal and right to left intracardiac shunt. Hemoptysis is a common complication of Eisenmenger syndrome and has been reported as the cause of death in 11-29% of patients. It can be caused by pulmonary artery thrombosis, pulmonary embolism, rupture of aortopulmonary collaterals, pulmonary artery dissection and hemorrhage due to an aneurysm or thin-walled arterioles, infectious sources or a bleeding diathesis. Treatment of hemoptysis in patients with Eisenmenger syndrome is challenging because they are at increased risk for bleeding and thrombotic complications. Hemoptysis in patients with Eisenmenger syndrome is often self-limited; however, it can be severe and life threatening. It is estimated that nearly 90% of patients with congenital heart disease survive into adulthood therefore adult pulmonologists may encounter this clinical scenario. Our patient’s hemoptysis resolved spontaneously and she remains clinically stable.

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