

Eisenmenger Syndrome Complicated by Pulmonary Artery Dissection

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Abstract

Keywords

- cardiac
- congenital heart disease
- CHD
- echocardiography (all modalities, applications)

This report describes a 40-year-old male patient diagnosed with pulmonary artery dissection involving both the left and right branches. The patient also had Eisenmenger syndrome secondary to an uncorrected patent ductus arteriosus. Diagnosis was facilitated through the use of transthoracic echocardiography and computed tomography. Due to the lack of definite guidelines for the optimal treatment strategy of this condition together with the poor condition of the patient, medical management was elected. Not surprisingly, the patient died suddenly only 7 days after the presentation.

A 40-year-old male patient with Eisenmenger syndrome secondary to an uncorrected patent ductus arteriosus (PDA) was admitted to the emergency room due to chest pain and dyspnea. On admission, oxygen saturation was 75% and blood chemistry analysis results were normal. An electrocardiogram showed left ventricular hypertrophy and thoracic radiographs confirmed the presence of cardiomegaly with pulmonary congestion. Transthoracic echocardiography (TTE) confirmed the presence of a large PDA with a right to left shunt (► **Fig. 1**). Computed tomography (CT) revealed dilatation of the main pulmonary artery and a dissection involving both the left and right pulmonary branches (► **Fig. 2**). The patient was treated conservatively and died suddenly 7 days after the presentation.

Discussion

Pulmonary artery dissection is a rare but life-threatening event in which the cause and pathogenesis are unclear. Pulmonary artery dissection is commonly associated with congenital heart disease accompanied by pulmonary hypertension. A PDA is the most frequent congenital heart disease associated with pulmonary artery dissection.^{1–4} It is reasonable to hypothesize that the high-flow rates in combination with secondary pulmonary hypertension associated with

a PDA predispose the pulmonary arteries to cystic medial necrosis. Histologic findings in patients with pulmonary hypertension have identified the presence of cystic medial necrosis in a significant number of cases, which is characterized by the extensive fragmentation of the elastin and collagen skeleton of blood vessels with pooling of mucopolysaccharides.⁵

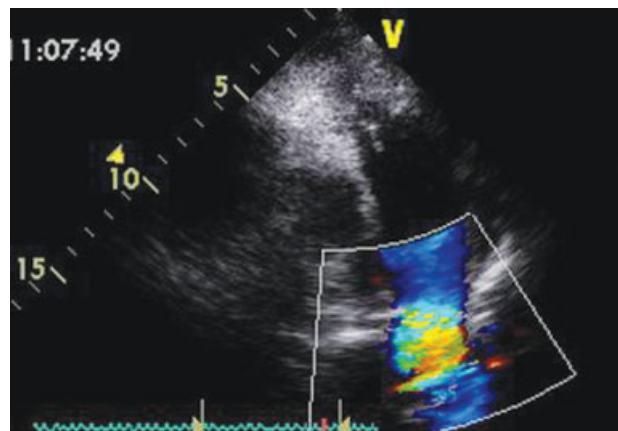


Figure 1 TTE showing a large patent ductus arteriosus with right to left shunt.

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Figure 2 CT showing dilatation of the main pulmonary artery and the dissection involved left and right pulmonary artery (arrow).

The symptoms of pulmonary artery dissection are non-specific. The most common symptoms are chest pain, dyspnea, cyanosis, hemorrhagic shock, acute pericardial tamponade, and sudden death. In the patient described herein, the clinical diagnosis was facilitated because of the

TTE and CT results. Nonetheless, the clinical decision regarding the treatment was difficult. Surgical repair seemed almost mandatory because of the exceedingly high risk of rupture of the dissecting, high-pressure pulmonary artery aneurysms; however, the chances of the patient surviving such an operation were considered very poor due to the Eisenmenger syndrome.

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