Pulmonary artery dissection in a patient with Eisenmenger syndrome treated with heart and lung transplantation

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Eisenmenger syndrome; Pulmonary artery dissection; Heart and lung transplantation.

Abstract We report the case of a patient with known Eisenmenger syndrome due to congenital ventricular septal defect, who developed pulmonary artery dissection. The patient was successfully treated with heart and lung transplantation. © 2003 The European Society of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

Ventricular septal defect is the most common congenital cardiac abnormality in infants and children. If not diagnosed in childhood the condition may lead to heart failure and pulmonary hypertension in adulthood. There is risk of sudden death partly due to malignant arrhythmias, but there have also been reports of pulmonary artery dissection diagnosed at autopsy. There have only been a few reports of pulmonary artery dissection diagnosed in vivo. The present report describes the successful treatment of pulmonary artery dissection by combined heart and lung transplantation in a patient with Eisenmenger syndrome due to congenital VSD.

Case report

A 39-year-old male with Eisenmenger syndrome due to a congenital VSD diagnosed at high age in a previously stable condition (NYHA class II—III) was admitted with acute chest pain, which was intensified by deep breathing. No coughing or dyspnoea was present. Before admission the patient was only receiving treatment with digoxin, 187.5 μg daily.
The patient was slightly cyanotic, but otherwise relatively unaffected. Blood pressure was 110/70 mmHg and heart rate 90 beats per minute. Auscultation of the heart revealed a crescendo systolic murmur grade IV and a decrescendo diastolic murmur grade II, most pronounced along the left sternal edge. Auscultation of the lungs was normal. There was no peripheral oedema.

Chest X-ray revealed an enlarged heart with prominent pulmonary vessels, but no signs of pulmonary oedema or infiltrates. ECG showed sinus rhythm, incomplete right bundle branch block, and right-sided hypertrophy. Transthoracic echocardiography showed a large VSD with bidirectional flow, overriding aorta, pronounced right side hypertrophy and dilatation, as well as dilatation of the common pulmonary artery. An abnormal structure was seen in the proximal part of the common pulmonary artery. Transoesophageal echocardiography confirmed the severely dilated common pulmonary artery (about 5 cm wide). The abnormal structure was identified as a dissection membrane starting 1 cm above the pulmonary valve cusps (Fig. 1). After acceptance from the patient he was evaluated for heart and lung transplantation. The time until transplantation was characterized by intermittent selflimiting supraventricular tachycardia and varying degrees of chest pain. The medical treatment consisted of increased doses of digoxin (250 μg BID), Felodipin 1.25 mg BID and diuretics, as well as analgetics. After 50 days of hospitalization he received combined heart and lung transplantation, and experienced a relatively uncomplicated postoperative course.

The dissection was confirmed during surgery and examination of the excised organs revealed pronounced atherosclerosis in the larger pulmonary arteries, and varying degrees of hypertensive changes in the peripheral pulmonary arteries, with intimal thickening, media hyperplasia and localized plexiform lesions. The patient was discharged from hospital two months after transplantation, and six years later the patient is well and works 60 h per week.

Discussion

VSD is the most common congenital cardiac abnormality in infants and children. Many defects close spontaneously within the first two years, but even up to 10 years of age spontaneous closure can be seen. Large defects give rise to heart failure during the first months of life and are, without surgical intervention, associated with high morbidity and mortality. Individuals with large defects who survive to adulthood often develop heart failure associated with pulmonary hypertension. With large

Figure 1  Transoesophageal echocardiogram through the aorta and the common pulmonary artery showing the dissection membrane (arrows).
defects the ventricular systolic pressures are equal, and the blood flow is determined by the difference in peripheral and pulmonary resistance (Eisenmenger syndrome). At this point surgical closure of the defect is not possible due to the risk of complications, and the condition is associated with risk of sudden death.

Acute spontaneous dissection of the main pulmonary artery is usually associated with high pulmonary artery pressure but also with connective tissue disease with normal pulmonary artery pressure. High pulmonary artery pressure is the result of characteristic pathological changes of the pulmonary artery tree, including intimal as well as medial thickening and occlusion of peripheral pulmonary arteries. In response to the increased pressure the main pulmonary artery undergoes pathological changes including atherosclerosis, mucoid degeneration, fragmentation of the elastic fibers and fibrosis. On this basis it is not surprising that pulmonary artery dissection can occur, and there have been several reports of fatal pulmonary artery dissection in patients with various forms of pulmonary hypertension including Eisenmenger syndrome. To our knowledge there has been only one report of surgical treatment for pulmonary artery dissection, and in that case it was in relation to primary pulmonary hypertension. Surgical treatment for Eisenmenger syndrome, either cardiac repair and single or dual lung transplantation, or heart—lung transplantation is reserved for advanced cases due to high perioperative mortality and poor prognosis. The choice of surgery for pulmonary artery dissection due to Eisenmenger syndrome is not established, but as we have shown, enbloc heart and lung transplantation is possible.

Conclusion

When encountering a patient with known pulmonary hypertension and acutely developed chest pain one has to be aware of the possibility of pulmonary artery dissection. Moreover enbloc heart and lung transplantation is a possible treatment of the disease.

References