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Abstract

Endocardial cushion defects are congenital abnormalities that result in valvular dysfunction as well as defects (or "holes") in the septa of the heart. They are typically diagnosed in early infancy; presentation late in life is rare. We present the case of a 72-year-old female admitted to the hospital with dyspnoea and palpitations. She was found to have multifocal atrial tachycardia. She suffered cardiac arrest associated with refractory hypoxaemia that required mechanical ventilation and vasodilator therapy with inhaled nitric oxide. Echocardiography revealed a large ostium primum atrial septal defect (ASD) complicated by Eisenmenger syndrome. It is likely that her arrhythmia, a sequela from her long-standing congenital abnormality, led to sudden decompensation. In this case presentation, we review the aetiology, presentation, and complications of ASDs.

Introduction

Atrial septal defects (ASDs) are common congenital cardiac abnormalities. They are classified as ostium primum, ostium secundum, sinus venosus defect, and coronary sinus defect [1]. Ostium primum is also referred to as an endocardial cushion defect. This occurs during development when the superior and inferior endocardial cushions fail to fuse. Endocardial cushion defects can result in abnormal formation of the atrioventricular (AV) valves and the interventricular septum.

Endocardial cushion defects are usually discovered at an early age. Echocardiography is the cornerstone of diagnosis [2]. Typically, a left-to-right shunt is present [1]. This leads to right heart dilation and a predisposition to atrial arrhythmias. The magnitude of the volume/pressure overload and the risk of complications increase with size of the defect and the presence of a shunt. Patients with large defects have a higher morbidity and mortality, highlighting the need for timely surgical repair.

Eisenmenger syndrome develops when the shunt is reversed from left-to-right to right-to-left as a result of pulmonary vascular disease [3,4]. It indicates that the pulmonary vascular disease is irreversible and surgical correction is contraindicated. This syndrome portends a poor prognosis. The case presented herein illustrates these points.

Case Report

A 72-year-old female presented with dyspnoea and palpitations. Prior to admission, she had experienced increasing exertional dyspnoea for several months. She was still able to complete her activities of daily living unassisted. Physical exam revealed an irregular tachycardia (rate 130 bpm), a holosystolic murmur at the left lower sternal border, a parasternal heave, and jugular venous distention. Chest radiography showed severe cardiomegaly and pulmonary vascular congestion. An electrocardiogram demonstrated multifocal atrial tachycardia and right ventricular hypertrophy. She was treated with diltiazem and the heart rate stabilized. Transthoracic echocardiogram showed a 2.3 cm septum primum ASD (Fig. 1), severe pulmonary hypertension, and a right-to-left shunt.

A few hours later, the patient sustained a cardiopulmonary arrest. Advanced cardiopulmonary life support was initiated and the patient was intubated. Spontaneous circulation returned after 15 min. She was transferred to the

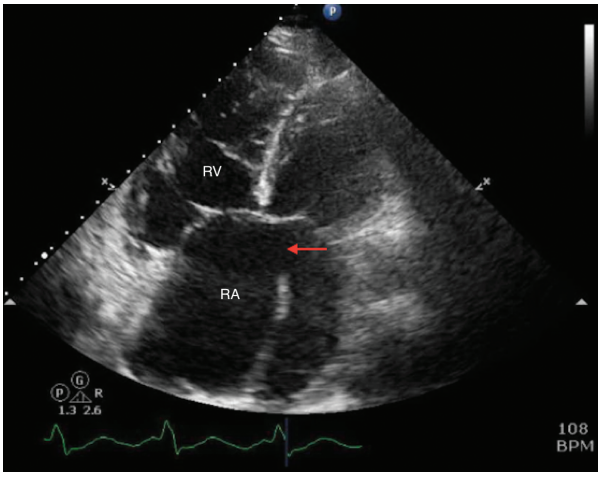

Figure 1. Apical four-chamber view from transthoracic echocardiogram showing a large septum primum atrial septal defect (arrow) and significant right atrial dilation.

cardiac intensive care unit but remained hypoxaemic despite a fraction of inspired oxygen (FiO_2) of 1.0. Right-to-left shunting was deemed to be the cause of persistent hypoxia. Inhaled nitric oxide was started at 20 parts per million. The oxygen saturations of the patient improved dramatically.

A transoesophageal echocardiogram revealed a septum primum defect with a now predominantly left-to-right shunt, an aneurysm of the perimembranous portion of the ventricular septum, and a single AV valve annulus but two separate orifices (Fig. 2). The presence of both a septum primum ASD plus a malformation in the AV valve annulus is consistent with a partial AV canal defect. A right heart catheterization demonstrated a pulmonary artery pressure of 61/6 mmHg and a pulmonary artery wedge pressure of 13 mmHg. Saturation study revealed a step-up in the oxygen saturations in the right atrium. The ratio of pulmonary to systemic blood flow (Qp/Qs) at the time of catheterization was calculated to be 3, confirming a left-to-right shunt. The catheterization was performed with the patient on inhaled nitric oxide.

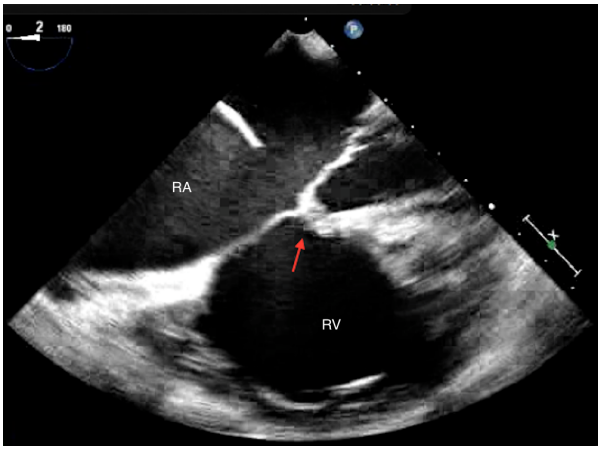

Figure 2. Transoesophageal echocardiogram demonstrating an aneurysm in the perimembranous interventricular septum (arrow) and a common atrioventricular annulus.

She was deemed a poor surgical candidate because of the presence of Eisenmenger syndrome. She was weaned off the nitric oxide and ventilator support. She was extubated on day 7, received a period of inpatient rehabilitation and was discharged home.

Discussion

This case highlights several key points: (1) congenital heart anomalies such as partial AV canal endocardial cushion defects rarely present in adults, especially in the seventh decade of life; (2) such defects can lead to fatal complications such as atrial tachyarrhythmias and even cardiac arrest; and (3) inhaled nitric oxide can be used as a salvage therapy in patients with refractory hypoxia and severe pulmonary vascular disease.

The exact aetiology of cardiac arrest in this case is unclear. There are several possible explanations. A fall in cardiac output due to a new onset atrial arrhythmia could have led to increased tissue oxygen extraction and a fall in the mixed venous saturation. In the setting of a right-to-left shunt, this would have led to a fall in arterial oxygenation further impairing cardiac output. Supplemental oxygen would have had little benefit in reversing hypoxaemia due to shunt. This spiral could have resulted in arrhythmia and cardiac arrest. Treatment with diltiazem may have led to hypotension and contributed to the decline as proposed above. Unfortunately, the patient was disconnected from telemetry monitoring in the minutes preceding the cardiac arrest so the mechanism of decline is speculative.

Long-standing partial AV canal endocardial cushion defects predispose to right heart dilation and atrial arrhythmias. With increasing pulmonary vascular resistance, the shunt is reversed from left-to-right to right-to-left. In this patient, a right-to-left shunt was discovered on the original echocardiogram. She sustained a cardiac arrest and refractory hypoxaemia. Inhaled nitric oxide, a vasodilator that activates cyclic guanosine monophosphate [5], was initiated as a salvage therapy. By decreasing the pulmonary vascular resistance, the effect of the right-to-left shunt was mitigated and oxygenation improved. A right heart catheterization and shunt study while receiving nitric oxide demonstrated an oxygenation step-up in the right atrium. Qp/Qs confirmed a left-to-right shunt. We speculate that the combination of inhaled nitric oxide and the

reversal of hypotension and hypoxaemia resulted in a return to the left-to-right shunt and her recovery.

Although the patient survived her hospitalization and dramatic presentation, her prognosis remains guarded due to the presence of pulmonary hypertension and advanced age. At the time of writing, she was scheduled for appointment in the pulmonary hypertension clinic for consideration of vasodilator therapy.

Disclosure Statements

No conflict of interest declared.
Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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