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Eisenmenger Syndrome in Adult Patient with Ostium Secundum Atrial Septal Defect

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Abstract

The diagnosis of congenital heart disease in adulthood is not unusual, but Eisenmenger syndrome is a rare complication associated with substantial morbidity and mortality. It occurs mostly in patients with post tricuspid defect or complex lesions. Exceptionally found in patients with pre-tricuspid defect, especially in the case of the ostium secundum atrial septal defect. While lung (plus defect repair) or combined heart and lung transplantation is thought be the definitive treatment for Eisenmenger syndrome, transplant organs are a limited resource and long-term results are still suboptimal. Eisenmenger syndrome patients have benefitted greatly from recent advances in the management of this condition, especially the introduction of pulmonary artery hypertension therapies. We report the case of a 39-year-old woman who was admitted with signs of progressive dyspnea who was diagnosed with ostium secundum atrial septal defect complicated by Eisenmenger syndrome.

Keywords: Eisenmenger syndrome; Pulmonary arterial hypertension; Atrial septal defect

Introduction

Congenital heart disease is the most common inborn defect, occurring in 0.8% of neonates [1]. Eisenmenger Syndrome (ES) is the most severe form of pulmonary arterial hypertension secondary to congenital heart disease with left to right shunt. Currently, this etiology has become rare in adults thanks to the technical development allowing an early diagnosis and thus a curative surgical treatment from the youngest age.

Case Report

A 39-year-old woman, mother of three children, without any prior medical history, presented with a history of exertional dyspnea evolving for 4 years, becoming stage III of NYHA classification accompanied by a cyanosis of the extremities in the 3 months preceding the consultation. Clinical examination found a eupneic patient at rest, its oxygen saturation was 95% at rest, but 78% at peak exercise. We note the presence of a generalized fingers clubbing. Her blood pressure was 120/85 mmHg, the heart rate at 70 bpm, without any sign of heart failure. An electrocardiogram revealed signs of right cavities enlargement. A chest radiograph revealed a cardiomegaly and a significant protrusion in the area of the pulmonary artery. Her haemoglobin level was 18.5 g/dl (hematocrit at 53%) with erythrocytosis (6.7 million/mm^3). Transthoracic echocardiography showed a dilation of the right cavities with atrial septal defect (type ostium secundum) with an estimated diameter of 26 mm. pulmonary artery dilated at 45 mm. moderate tricuspid regurgitation with severe pulmonary arterial hypertension.

The estimation of pulmonary vascular resistance (V_{max} of tricuspid regurgitation/pulmonary velocity time integral) was at 0.54 (normal ≤ 0.2). Subsequently, we performed transesophageal echocardiography that revealed a 26 mm ostium secundum atrial septal defect with bidirectional shunt (Figure 1) and confirmed the severe pulmonary artery hypertension (systolic pulmonary arterial pressure at 135 mmHg. A cardiac catheterization revealed a mean pulmonary arterial pressure at 82 mmHg with high pulmonary vascular resistances=10.4 UI Wood superior to 2/3 of systemic vascular resistances (12 UI Wood) without improvement after O_2 administration (10 liters for 10 minutes). The angiographic computed tomography (Figures 2 and 3) eliminated aorto-pulmonary fistula or other abnormality and demonstrated the dilation of pulmonary artery and its branches as well as right heart cavities (Figures 4A and 4B). The patient was treated with Bosentan (125 mg/day) combined with a diuretic (furosemide 40 mg/day+spironolactone 75 mg/day).

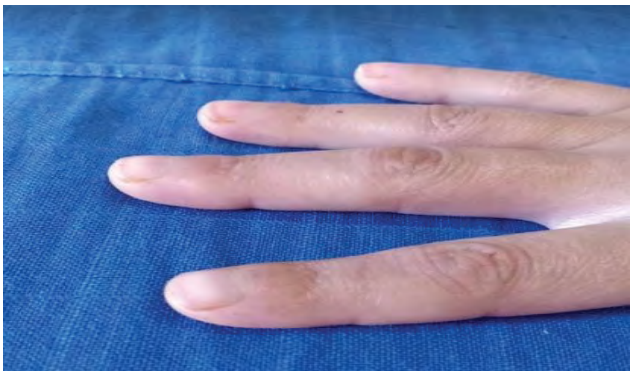

Figure 1: Fingernails clubbing.

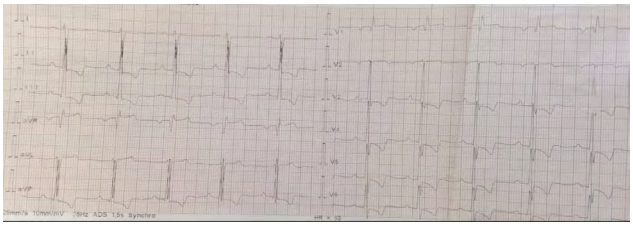

Figure 2: Electrocardiogram showed signs of right cavities enlargement.

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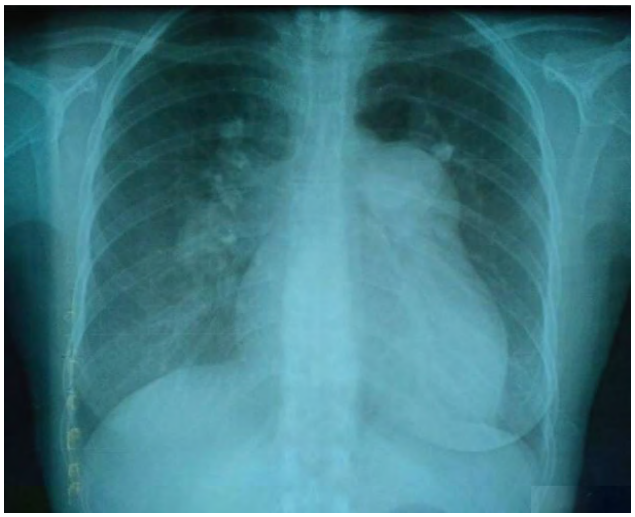

Figure 3: Cardiomegaly and a significant protrusion in the area of the pulmonary artery at chest radiograph.

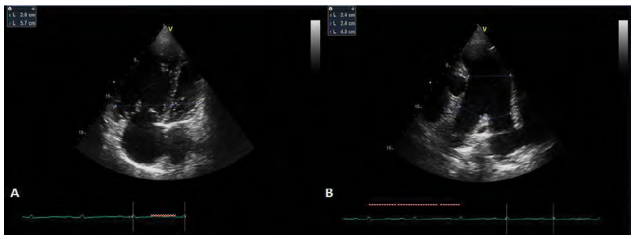

Figure 4: TTE showed a dilation of the right cavities with ostium secundum atrial septal defect (A) and pulmonary artery dilation at 45 mm (B).

Discussion

There are a multitude of very different clinical pictures of patients with PAH associated with CHD (PAH-CHD). However, four broad clinical phenotypes have recently been defined: Eisenmenger syndrome, PAH associated with systemic-to-pulmonary shunts, PAH associated with small defects, and PAH after cardiac defect correction [2]. Eisenmenger syndrome groups all the intracardiac or extracardiac communications that begin by systemic pulmonary shunts and evolve towards a severe increase in the pulmonary vascular resistance and shunt reversal (pulmonary to systemic or bidirectional), characterized clinically by exercise intolerance and chronic cyanosis [3]. The group with extracardiac communication included persistent ductus arteriosus, aortopulmonary windows, large Waterston or Pott's anastomosis (Figures 5A-5C).

The exact prevalence of ES is not known. According to historical data and before the advent of timely intervention, 8% of patients with congenital heart disease and 11% of those with left-to-right shunts developed Eisenmenger syndrome [4]. Eisenmenger syndrome is a major complication of shunt lesions, especially post-tricuspid and complex shunts, more rarely pre-tricuspid shunts including atrial septal defects. Thus, in non-operated patients, only 10% with large atrial septal defect are at risk of developing Eisenmenger syndrome, compared to about 50% of wide ventricular septal defect [3]. In the cohort of Kempny et al. [5] include 1098 adult patients c syndrome, only 140 (12.7%) had a pre-tricuspid defect. This minority of patients with pre-tricuspid shunt seem to be afflicted both from the cardiac lesion and from predisposition, maybe genetic. The occurrence of fixed pulmonary hypertension, complicating an ostium secundum atrial septal defect, is rare (<8%) but more frequent in women and favored by multiple pregnancies [6]. As the case for our patient who has been pregnant for three times (Figures 6A-6D).

At the end of their study, Kempny et al. concluded five simple, non-invasive predictors of death in this population, namely age, underlying shunt type, oxygen saturation, presence of sinus rhythm and presence

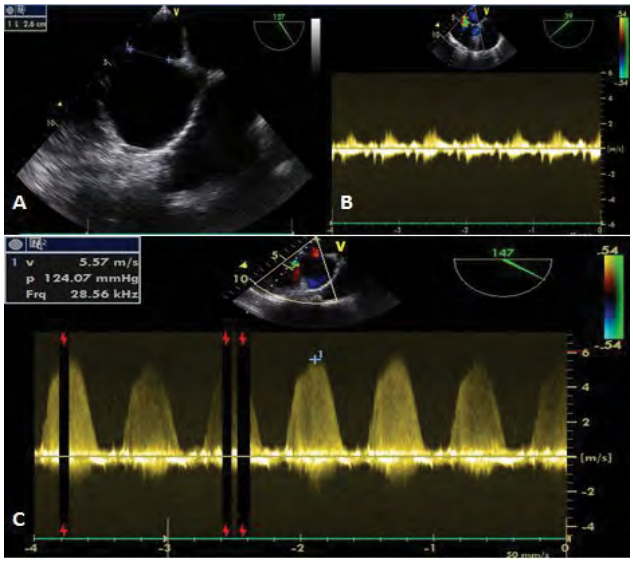

Figure 5: Transesophageal echocardiography revealed ostium secundum atrial septal defect measuring 26 mm (A) with bidirectional shunt (B) and severe pulmonary artery hypertension (systolic pulmonary arterial pressure at 135 mmHg) (C).

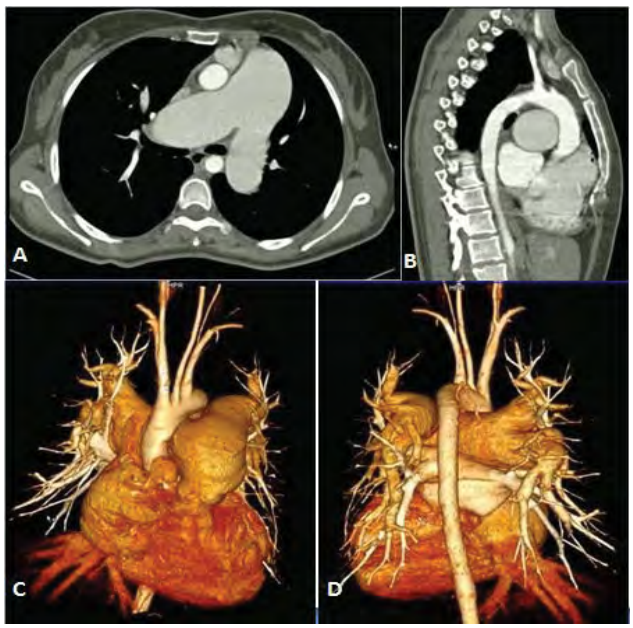

Figure 6: Angiographic CT demonstrated a dilated pulmonary artery and right cavities in axial section (A), sagittal section (B) and 3D reconstruction (C and D).

of pericardial effusion [5]. Fortunately, our patient has none of these aforementioned factors. Other factors related to death have been described in other studies, such as the cardiac failure and poor NYHA functional class [7-9], low body weight and low body mass index [6] and renal dysfunction [10]. At this stage of evolution, the defect cannot be repaired, but the therapeutic advances concerning the specific pulmonary artery hypertension drugs letting hope the improvement of the symptoms, the hemodynamic data, the quality of life and the survival [3]. Mariana et al. demonstrated a direct correlation between longer exposure to these specific drugs and better outcomes in patients with Eisenmenger syndrome [11]. The only definitive treatment for Eisenmenger syndrome is lung transplantation with shunt closure or heart and lung transplantation, but transplant organs are a limited resource and long-term results are still suboptimal.

Conclusion

Eisenmenger syndrome should disappear thanks to the early diagnosis of left-right shunts. However, there will be always patients who escape from this step diagnosis either by evaluation "error" or by their geographical origin. These patients have a much better prognosis than other cases of pulmonary hypertension, but have a pejorative functional prognosis related to extreme cyanosis and the erythrocytosis resulting from it. There is currently no curative treatment and the only way out from therapeutic escape is cardiopulmonary transplantation whose results are very disappointing. Prevention and early closure of the shunt must be a priority because they allow healing.

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