Pulmonary Embolism or Eisenmenger Syndrome?

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Introduction
Eisenmenger syndrome is a late complication of untreated congenital heart disease associated with left to right shunt, that can prompt in situ pulmonary thrombosis. Herein the case of a 41 year old man with suspected pulmonary embolism, where an atrial septal defect lead to Eisenmenger Syndrome, is presented. Only lung transplantation and surgical defect closure revealed to be the optimal treatment.

Case Report
Herein a case of a 41 year old male, healthy until the age of 39, is presented. He started to refer NYHA functional class II dyspnoea, cough and peripheral edema in December 2011. As his condition worsened, he was admitted to a suburban Emergency Department, where a thoracic CT-scan showed massive bilateral pulmonary thrombosis and a diagnosis of subacute pulmonary embolism was made. Thrombophilic screening, abdomen CT-scan, HIV screening, venous Doppler and tumoral markers were negative. The patient was discharged on oxygen therapy and was referred to our pulmonary hypertension unit for clinical evaluation and follow up.

At first evaluation the patient was symptomatic for dyspnoea class NYHA II and saturation at rest in room air was 80%. Systemic blood pressure was 120/80 mmHg. Clinical examination showed hepatomegaly and peripheral edema. ECG showed sinus tachycardia (110 bpm) and incomplete right bundle branch block. Transthoracic echocardiography showed normal left ejection fraction (EF 60%), severe dilatation of the right ventricle (lateral wall thickness 8 mm, end diastolic volume 260 ml, TAPSE 19 mm, FAC 32%); indirect estimation of the systolic pulmonary arterial pressure was at a value of 90 mmHg. Blood analysis showed polycythemia (Hb 18.4 g/dl, Hct 57%) and hypoxemia in room air (artery pO2 57 mmHg). Thoracic CT-scan was repeated and showed severe pulmonary artery dilatation (trunk diameter 51 mm) with hypodense appositions in the right pulmonary main artery and all its lobar branches (Fig. 2), in the main left pulmonary artery and in the left superior lobar artery. The initial diagnosis of subacute pulmonary embolism was confirmed and the patient was hospitalized in our Intensive Care Unit.

Control of heart failure was attempted with non-invasive ventilation, diuretics, nifedipine and nitroprusside, without

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Highlights
Eisenmenger syndrome is a late complication of untreated congenital heart disease associated with left to right shunt, that can prompt in situ pulmonary thrombosis. Herein the case of a 41 year old man with suspected pulmonary embolism, where an atrial septal defect lead to Eisenmenger Syndrome, is presented. Only lung transplantation and surgical defect closure revealed to be the optimal treatment.

Keywords: Eisenmenger Syndrome; Congenital heart disease; Pulmonary thrombosis

improvement. Therefore, transesophageal echocardiography was performed and demonstrated the presence of an ostium secundum type atrial septal defect, with a diameter of 20 mm and bidirectional shunt. The association of CHD, pulmonary thrombosis, cyanosis and severe pulmonary hypertension lead to a diagnosis of Eisenmenger Syndrome in a patient with ASD. Right catheterization showed pulmonary hypertension and bidirectional interatrial shunt, with systolic aortic pressure (SAP) of 90 mmHg, an average pulmonary arterial pressure (PAP) of 50 mmHg, wedge pressure of 5 mmHg, pulmonary vascular resistances (PVR) of 15.63 UW and a QP/QS ratio of 1.1. Treatment with endothelin-inhibitor (bosentan) and anticoagulation was started and well tolerated, and led to gradual clinical improvement.

Clinical evaluation and previous examinations were repeated in September 2012. Echocardiography remained unchanged. Blood gas analysis (BGA) in room air confirmed severe hypoxemia (pO2 48 mmHg). Right catheterization confirmed a severe pulmonary hypertension and severe increased pulmonary resistances (SAP 110 mmHg, PAP 50 mmHg, wedge pressure 7 mmHg, RVP 8.94 UW, QP/QS 1).

Considering both clinical conditions and persistent pulmonary hypertension, sildenafil was associated to bosentan, and was well tolerated. A check up at three months on dual therapy showed that echocardiography and right catheterization were unchanged and thoracic CT-scan evidenced thrombotic appositions only in the left pulmonary arteries.

Considering clinical and instrumental data, the patient was accepted on the lung transplantation waiting list (December 2012). Six months later, on 26th June 2013, the patient had bilateral lung transplantation and ASD surgical closure.

The post surgery period was complicated by an A2 reject, treated with corticosteroids, and anastomotic bronchial stenosis, which was treated with endoscopic bronchial stenting. At 3 months post lung transplantation, the patient was asymptomatic for dyspnoea and showed no signs of heart failure. Echocardiography showed a normal left ventricular function (EF 60%), no residual interatrial shunt, hypertrophy and normal dimensions and function of the right ventricle (TAPSE 20 mm, FAC 40%); PAP was 25 mmHg.

Figure 1. Echocardiography - Apical 4 chambers view shows severe right ventricle dilatation.

Clinical and echocardiographic stability was confirmed one year post lung transplantation.

**Discussion**

Eisenmenger Syndrome occurs when a CHD with left-to-right shunt prompts persistent exposure of pulmonary vasculature to increase blood flow and lead to a pulmonary obstructive arteriopathy and a pulmonary vascular resistance close to, or above, that of systemic vascular resistance. The original left to right shunt reverses and turns into either right to left or bidirectional shunt. Dyspnoea, low arterial saturation, cyanosis, compensatory polycythemia, blood hyperviscosity and capillary damage occur and may result in fatigue, syncope, cerebrovascular accidents, brain abscesses, or even sudden death. Hemoptysis may occur as a result of lung infarction, rupture of dilated pulmonary arteries, thrombocytopenia, platelet function disorder or clotting factor deficiencies.

The risk of thrombosis is also enhanced by the coagulation disorders and polycythemia. The fibrinolytic system is impaired by blood stasis due to bronchial arterial hypertrophy and dilation, endothelium damage and the activation of procoagulant factors [1]. CT scan is able to detect in situ pulmonary thrombosis, which is quite a common finding in Eisenmenger Syndrome, with a reported incidence of around 21-29% [2].

Ventricular septal defect (VSD) is the most frequent underlying cause, ASD rarely evolves into Eisenmenger syndrome [3]. The treatment regime to be adopted for pulmonary hypertension in Eisenmenger syndrome is mainly based on clinical experts consensus [4]. Long term oxygen therapy is able to reduce dyspnoea and, although there is a lack of literature support, specific drugs such as endothelin receptor antagonists, phosphodiesterase type-5 inhibitors and prostacyclin analogues, may be indicated for pulmonary hypertension [5,6,7,8]. Management of concomitant...
bleeding and clotting increased risk is controversial. There is a paucity of widely accepted indications and anticoagulant therapy management of patients with Eisenmenger Syndrome must be tailored. The option of surgical treatment with thrombectomy and aneurysm repair in the presence of pulmonary thrombosis is a high risk solution and pneumonectomy is not recommended [9,10].

Surgical correction of the primitive defect in Eisenmenger Syndrome is not recommended, therefore in most of patients symptomatic therapy is the only therapeutic chance. Lung transplantation with correction of CHD or heart and lung transplantation can be performed in selected patients.

**Conclusion**

The case described herein reports a patient with pulmonary thrombosis in an undiagnosed congenital ASD, which led to a preliminary diagnosis of right heart failure due to subacute pulmonary embolism. Anticoagulation and medical treatment for pulmonary hypertension were started once the diagnosis had been corrected. However, as severe pulmonary hypertension and bidirectional shunt persisted despite medical therapy, lung transplantation and surgical ASD correction was the only therapeutic choice left.

This report emphasizes the importance of taking into consideration undiagnosed CHD in adults with pulmonary hypertension and pulmonary thrombosis, because of serious clinical consequences.

**Declarations of Interest**

The authors declare no conflicts of interest.

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**Legend**

ASD: Atrial septal defect  
BGA: blood gas analysis  
CT-scan: computed tomography  
EF: ejection fraction  
FAC: fractional area change  
Hb: hemoglobin  
Hct: hematocrit  
NYHA class: New York Heart Association functional class  
SAP: systolic aortic pressure  
PAP: average systolic pulmonary arterial pressure  
pO2: partial pressure of oxygen  
PVR: pulmonary vascular resistences  
Qp/Qs: pulmonary flow/systemic output  
TAPSE: tricuspid annular plane systolic function  
VSD: Ventricular septal defect

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