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A 22-Year-Old Woman With Eisenmenger Due to Atrial Septal Defect; A Case Report

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Abstract

Atrial septal defects (ASDs) constitute a common form of congenital heart defect (CHD) and can lead to significant morbidity and mortality in adulthood if left untreated. While advancements in prenatal screening and fetal echocardiography have facilitated early interventions, some cases may go undetected until adulthood, manifesting as symptoms like progressive shortness of breath. A recent case involved a 22-year-old woman with a history of ASD since childhood, presenting with cyanosis, digital clubbing, and peripheral edema. Examination revealed cardiomegaly, right atrial enlargement, and right ventricular hypertrophy. Diagnostic tests, including chest X-ray, electrocardiogram, and echocardiography, confirmed features consistent with Eisenmenger's syndrome, characterized by severe pulmonary hypertension and a large secundum ASD. Effective management requires a multidisciplinary approach involving cardiologists, pulmonologists, cardiac surgeons, and internists. It is crucial to address the underlying causes, as medications targeting pulmonary vessels may not consistently provide reliable results. Timely intervention and collaboration among specialists are paramount in improving outcomes for individuals with ASDs and preventing complications associated with Eisenmenger's syndrome.

Keywords: Eisenmenger Syndrome, Pulmonary Arterial Hypertension, Atrial Septal Defect

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INTRODUCTION

A 22 year-old-woman was admitted with a slowly progressing shortness of breath. This symptom became more frequent after cesarean delivery five months ago. Malaise and cough has been present since several weeks ago. Patient was brought by her family and during anamnesis, patient does not respond to questions and appears to look away. She had reported for having ASD since childhood, with no cardiac surgical history before. She received antituberculosis drugs for six months last year. Physical examination revealed an SpO2 of 77% at room air BP of 112/84, no increase of jugular venous pressure, peripheral and central cyanosis, digital clubbing and pitting edema peripheral. On respiratory examination, there were bilateral rales sounds on all lung fields and respiratory rate (RR) of 36 on rest. On cardiovascular examination showed loud P2 with no murmur. The patient was placed on non-rebreathing mask oxygen therapy with improvement of oxygen saturations to 90-94%. Laboratory examination showed: Hemoglobin 16,9 g/dl, Hematocrit 61,8%, Trombocyte 216.000/ul, MCH 22,6,

MCHC 27,3. The ureum was elevated to 62,4 mg/dl (normal level 12-42mg/dl) and eGFR showed 94,6 mg/min/m2. Blood gas analysis show decreasing pO2 47mmHg and SatO2 81,6%. The natrium level was decreased to 128,1 mmol/L and Chloride 91,7%. Chest X-ray on the emergency unit showed cardiomegaly with right atrial enlargement and right ventricular hypertrophy, pulmonary hypertension features, increasing bronchovascular, infiltrate in both upper lungs. (Figure 1) Urgent electrocardiogram confirmed sinus rhytm, HR 100 bpm, right axis deviation, right ventricle hypertrophy and incomplete RBBB. (Figure 2) Patient began to be treated in ICU and given furosemide drip 10 mg/hour, sildenafil 20 mg 3x1, spironolactone 25 mg 1x1. Echocardiography showed left ventricular hypertrophy, LV D-shape, dilatation of the right atrium and ventricle, ejection fraction 54%, global left ventricle normokinetics, severe tricuspid regurgitation TVG 83 mmhg, severe pulmonary hypertension, ASD secundum in diameter 31-33 mm. This patient was consulted to pulmonologist with suspicion of relapse of TB, chest ultrasound was performed and demonstrated left pleural effusion. Intravenous antibiotic ceftazidime 3x1 gr was started, and patient was given inhalation ventolin and pulmicort 2x1.

On day two, albumin test was ruined and showed hypoalbumin: 2,33 g/dl. Treatment was administered: Albumin 20% 100 cc and albumin capsule 3x2. Patient still quiet when asked by physician and then consulted to psychiatrist for further evaluation.

On day three, complaints of shortness of breath reduced, gradual O2 tapering from NRM 10 lpm to 8 then 6 lpm with SpO2 target above 80%. The patient respond to questions with meaningless sounds, nods and blinks. When asked about feelings of sadness the patient nodded and cried. According to her father, the patient was divorced 6 months ago and tends to silent and keeps to herself. No history of previous psychiatric treatment. Patient was diagnosed with depression and start treatment sertraline 1x12,5 mg and psychotherapy.

On day four, complaints of shortness of breath are decreasing, swelling in the legs was resolved, SpO2 80% on oxygen device that been replaced with a 4 lpm nasal cannula, furosemide drip therapy has decreased the dose to 3 mg/day. Digoxin 0,125 mcg 1x1 was administered on day five and furosemid switch into 1x 20mg intravenous injection. Antibiotic was stopped on day six, patient was planned to discharged the next day because complaints of shortness of breath had improved, and SpO2 showed 85-86% room air. Oral therapy given: furosemide 40 mg 1x1/2, digoxin 0,225mg 1x1, sildenafil 20 mg 3x1 and albumin capsule 3x2.

During outpatient control, a new complaint was occurred, the right hand and right leg could not be moved for 2 days, the patient was consulted to a neurologist for further evaluation.

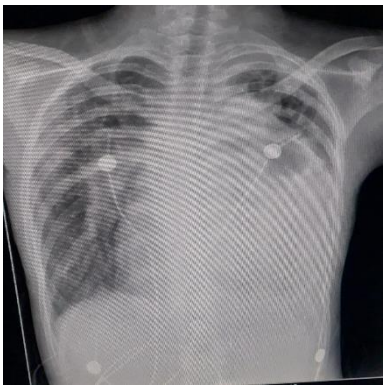

Figure 1. Chest X-Ray demonstrated right atrial enlargement and right ventricular hypertrophy, pulmonary hypertension features, increasing bronchovascular, infiltrate in both upper lungs

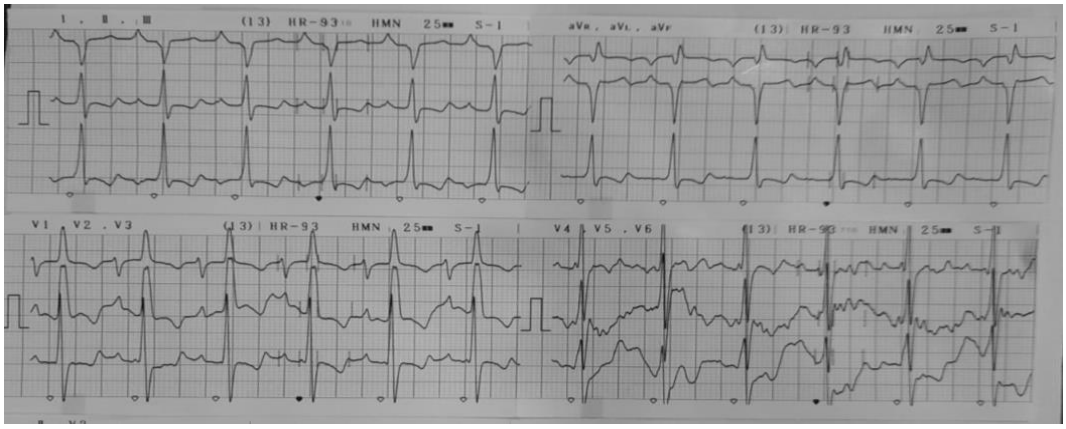

Figure 2. Electrocardiogram demonstrated normal sinus rhytm, right axis deviation, right ventricle hypertrophy and incomplete RBBB

RESEARCH METHOD

This study is a case report which detailed description of patients and clinical case with a disease and complication, uncommon combination of diseases. It can also describe new or inovative approach to the diseases.

RESULTS AND DISCUSSION

Eisenmenger syndrome is the most severe pulmonary arterial hypertension phenotype linked associated with congenital heart disease. The most frequent presentation is a patient with previously diagnosed congenital heart disease (CHD) such as ASD, VSD and PDA defects. While CHD are frequently correctable, failure to do so may result in Eisenmenger syndrome.4 Patients initially develop left-to-right shunting with volume overload of the pulmonary circulation due to the predominant distribution of vascular resistance between pulmonary and systemic circulation, frequently accompanied by elevated pressure due to post-tricuspid shunt lesions. Pulmonary vascular disease develops and worsens over time to the point where shunt reversal takes place, and it first appears during exercise before later appearing at rest.2 Three main processes ultimately result in the conversion of a left-to-right shunt into a right-to-left shunt:

imbalance in pulmonary vascular tone that causes vasoconstriction, fibrosis, the proliferation of pulmonary vascular smooth muscle that results in vascular remodeling, and an increased blood flow resistance that causes thrombosis. This mechanism has been associated to changes in the expression of vasoactive mediators such as endothelin-1, thromboxane, prostacyclin, and nitric oxide.5

The characteristic features of ES include multiorgan involvement, chronic hypoxemia, secondary erythrocytosis (often associated with iron deficiency), a high burden of arrhythmias, an increased risk of infection, progressive heart failure (HF), also have the highest rate of renal impairment among CHD patients.1 In our case, the patient had signs of hypoxemia such as shortness of breath at rest, central and peripheral cyanosis, clubbing fingers and the presence of secondary erythrocytosis which was demonstrated from laboratory findings of increased hemoglobin, hematocrit and erythrocytes. Clinical follow-up should prioritize routine monitoring of annual complete blood count, iron tests, kidney function, and uric acid levels in order to treat any anomalies that may emerge. Secondary erythrocytosis is advantageous for adequate oxygen transport and delivery. Continuous supplemental oxygen is indicated when the arterial blood oxygen pressure is regularly 60 mmHg44, except in Eisenmenger patients, when it is only recommended if it results in a demonstrated, consistent, and significant rise in oxygen saturation and symptom relief.6 An improvement in symptoms and increasing oxygen saturation were seen in this patient during treatment in the ICU. Severe pulmonary vascular disease with ES is very rare and seems to be related to age, size of the defect, female sex, and non closed ASD status and may be seen in those with genetic predispositions, like in idiopathic PAH.6 ASD recognition and correction early after childbirth has helped reduce the incidence of PAH. In the study by Forlemu et al7, patients with ASD ES had significantly larger ASD (2,5×3,5 cm) with secundum type defect. The patient in our case occured in her 2nd decade with PAH ES and a big ASD (31-33 mm), implying that the magnitude of the ASD likely played a significant hemodynamic role in the reversal of the shunt and the development of ES.

(extracardiac) Non-cardiac comorbidities may become increasingly crucial with ongoing age. Patients tends to having psychiatric comorbidities were younger at the time of hospitalization compared to those having no psychiatric illness. Anxiety (12.1%) and depression (11.4%) were the most common psychiatric comorbidities. Neidenbach et al' studies showed psychiatric disorders (52/821, 6.3%) are very common as they may considerably influence the cognitive and psychosocial development of CHD patients. Additional etiologic factors are cerebral hypoperfusion and embolism, in part induced by cardiopulmonary bypass and cardiac arrest in accidental hypothermia. It is known that depression has a negative influence on the cardiovascular function, morbidity and mortality. The neglection of medical advice in terms of medication and life-style, has significantly increased the risk of a recurrent cardiac event and a higher mortality. Neurological problems are important sequelae in ACHD. In a retrospect analysis of more than 23,000 CHD patients, Hoffmann et al. demonstrated an incidence of cerebrovascular accidents in more than 2%. Cyanotic patients, with a high rate of ischemic stroke and consecutive longtime disability, are especially

affected. As in our case, patients patient experienced depression and early symptoms of stroke which needs to be further evaluated.

Therapy for pulmonary hypertension includes general supportive, assessment of vasoreactivity, and administration of vasoactive medications. Five classes of drugs target three major cellular signaling pathways: the endothelin pathway, the nitric oxide/cyclic guanosine monophosphate (cGMP) pathway, and the prostacyclin pathway.(guideline) The significance of sequential PAH therapeutic technique in Eisenmenger syndrome, as well as the utilization of 6MWT for therapy decision-making. In Eisenmenger patients with impaired exercise capacity (6MWT distance 450 m), an initial endothelin receptor antagonist monotherapy followed by combination therapy should be tried if patients do not improve. According to ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension 2022, Bosentan is recommended in symptomatic patients with Eisenmenger syndrome to improve exercise capacity. Bosentan improved 6MWD and decreased PVR in patients with Eisenmenger syndrome in WHO-FC III. Meanwhile, moderate- to high-risk condition and requires a proactive approach using initial or sequential combination treatment, including parenteral prostacyclins. Sildenafil and tadalafil (Phosphodiesterase type 5/PDE5's group) have shown favourable functional and haemodynamic results in patients with PAH-CHD and Eisenmenger syndrome. PDE5 is the major enzyme responsible for cGMP degradation in pulmonary vascular smooth muscle, and increased in patients with PAH. It delay the metabolism of cGMP and thereby potentiate the vasodilatory efects of NO and the natriuretic peptides. They also cause smooth muscle relaxation and vasodilation by increasing cGMP levels, by increase cGMP synthesis. Shadaab et al, reported a significant improvement in the 6 MWD and oxygen saturation in the monotherapy group and both parameters had insignificant improvement in the combination therapy group. The NYHA functional class had significant improvement in both groups and mean pulmonary artery pressures significantly decreased in the combination therapy group.

Surgical correction of the underlying heart defect is generally not recommended. In patients who have developed pulmonary arterial hypertension (PAH) as a consequence of unrepaired congenital heart disease (CHD), the defect itself may act as a protective measure, preventing the worsening of pulmonary vascular resistance in the face of increasing right ventricular pressure.

Patients diagnosed with Eisenmenger syndrome are likely to have a shorter life expectancy, with increased mortality in their third and fourth decades. Common causes of death include ventricular failure, hemoptysis, pregnancy complications, and strokes. In individuals experiencing Eisenmenger syndrome, it is recommended to avoid certain scenarios such as pregnancy, dehydration, isometric exercise, iron deficiency anaemia, and prolonged time spent at high altitudes. When prescribing antihypertensive medication, caution should be taken with the use of peripheral vasodilating agents, as they may exacerbate the right-left shunt. Due to in-situ thrombosis and reactive erythrocytosis, individuals diagnosed with Eisenmenger syndrome may display indications of both a bleeding tendency and a prothrombotic state.

CONCLUSION

Eisenmenger's syndrome is best managed by a multidisciplinary team that includes a cardiologist, a pulmonologist, a cardiac surgeon, an internist. There are many causes of the disorder and the key is to treat the primary disorder. Medications that widen the pulmonary vessels are not reliable or consistent in their effectiveness.

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