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Case Report

Eisenmenger syndrome in a patient with ventricular septal defect: a case report

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ABSTRACT

Eisenmenger Syndrome (ES) represents Pulmonary Arterial Hypertension (PAH) associated with Congenital Heart Defects (CHD). Although patients survive until their third or fourth decades of life, the symptoms include dyspnea, cyanosis, fatigue, dizziness, and syncope. In addition, cardiac arrhythmias, a late complication are causing sudden death in patients with ES. Treatment options have been limited; however, recent successes have been achieved with the use of therapies targeted against the pathophysiological pathways that underlie PAH. The dual endothelin receptor antagonist and prostacyclins demonstrated to improve hemodynamics of the patients. This is the case of a 16 year old young female with ventricular septal defect that was admitted with increasing shortness of breath and cyanosis with clubbing which are clinical features of Eisenmenger syndrome. She was medicated with Furosemide, Sildenafil which improved her functional status.

Keywords: Pulmonary hypertension, Eisenmenger syndrome, Sildenafil

INTRODUCTION

Eisenmenger syndrome is an untreated congenital cardiac disorder with intracardiac communication that leads to dyspnea, pulmonary hypertension due to reversal of flow, and cyanosis.[1-3] This was observed by Viktor Eisenmenger in 1897 in a 32 year old male who had dyspnea and cyanosis since childhood and died with massive hemoptysis. Later, Paul wood described it as Eisenmenger complex, consisting of pulmonary hypertension with reversed or bidirectional shunt or a ventricular septal defect.[1] Eisenmenger syndrome is commonly develops before puberty but is also observed in adolescence and early adulthood. Patients in underdeveloped countries more possibly present with late undiagnosed and uncorrected congenital cardiac lesions with a distinct elevated Pulmonary Vascular Resistance (PVR). They are probably becoming untreatable secondary to Eisenmenger physiology. These patients require long-term tertiary care and 5% to 10% of them present Pulmonary artery hypertension (PAH) that affects morbidity, mortality and quality of life.[4] We report the case of a 16-year-old female who presented with lower extremity edema, cyanosis of the extremities and ventricular septal defect, fulfilling the criteria for ES.

CASE REPORT

A 16 year old female patient was admitted with signs of shortness of breath for 2 days, pleuritic chest pain with non-productive cough and severe edema of the lower extremities and cyanosis of the fingertips. Medical history recounts Ventricular Septal Defect (VSD) and on treatment. Family history revealed nothing significant with respect to heart disease. On clinical examination, she was moderately built and nourished. General examination revealed edema, clubbing and central cyanosis. Her vitals were stable with a pulse rate of 110 beats/minute and

blood pressure 120/100 mm of Hg in the right upper limb. Her respiratory rate was 24/minute. Thyroid, breast and abdominal examination revealed no abnormality. On cardiovascular auscultation a loud second sound, an ejection click with diastolic murmur in the left parasternal region. Lungs were clear with normal bilateral vesicular breath sounds. On investigation, hemoglobin was 9 g/dl and hematocrit was 65%. Complete blood count and urine examinations were found to be normal. Chest radiograph showed enlargement of the right atria and cardiomegaly (Figure 1), especially right-sided enlargement. The ECG showed features of ventricular hypertrophy with at all R wave in leads VI and II and deep S wave in lead V (Figure 2) and echocardiography showed an increase in the ventricle with significant tricuspid regurgitation, enlarged right ventricle with Left Ventricular Ejection Fraction (LVEF) 47%. The patient was started with high flow oxygen and tab. Furosemide 20 mg OD, tab sildenafil 20 mg OD, tab acitron 2 mg OD and cap augmentin 625 mg BD for infective enocarditis prophylaxis. The patient was discharged after 5 days with a progressive improvement of functional status.

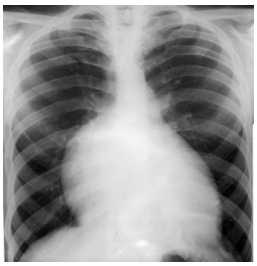

Figure 1: X-ray showing enlargement of the right atrium.

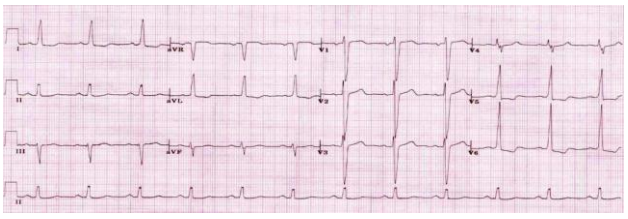

Figure 2: ECG pattern with Bi ventricular hypertrophy.

DISCUSSION

Eisenmenger's Syndrome (ES) is a rare complex combination of cardiovascular abnormalities defined as pulmonary hypertension with reversed or bi-directional shunting through an intracardiac or aortopulmonary communication. Its exact incidence is unknown. [1] This syndrome generally develops before puberty, but also been observed in adolescence or early adulthood. [5] It affects both males and females. Infants born with a ventricular septal defect or atrial septal defect or both, or a persistent patent arterial duct are at high risk of developing ES. Clinical manifestations include dyspnea (especially with exertion) cyanosis, chest pain, lethargy, syncope, fatigue, dizziness, palpitations, arrhythmias, and, not often, right heart failure (associated with hepatomegaly, peripheral edema, jugular venous distention). Fingers clubbing and cardiac murmurs may occur. Hemoptysis is a late symptom. Cardiac failure and sudden death may also occur.[2] Laboratory testing shows increased hematocrit value. Electrocardiogram shows majorly right ventricular hypertrophy and, occasionally right atrial hypertrophy. In ES there will be communication between two cardiac chambers, either due to congenital cardiac malformation or by a surgically-created shunt, which allows oxygenated blood to return back into the right ventricle and to the lungs, causing an increase in pulmonary blood pressure. The diagnosis is based on the clinical manifestations and on the abnormal electrocardiogram recordings and clinical imaging.[7] Cardiac catheterization, for measuring pulmonary arterial pressures, confirms the diagnosis. Management includes avoiding conditions that may exacerbate the syndrome, such as pregnancy, dehydration, exercise, travel, planes, and climbing high altitudes.[8] The syndrome can be avoided by correcting the septal defects by surgical procedure prior to permanent changes within the lungs. Medical treatment is supportive. The pulmonary hypertension may be treated with prostacyclin and endothelin antagonists. Prophylaxis against endocarditis is recommended.[9,10] There is no specific treatment, apart from both heart and lung transplantation: Particularly for those with a poor prognosis and fail to respond to medical therapy. The patient described has a rare disease with atypical presentation, considering that age is not typical of the disease or the response to treatment with Furosemide, Sildenafil and Augmentin, achieving a good outcome.

CONCLUSION

Treatment with drugs like Sildenafil and Furosemide will improve the prognosis and quality of life of those cases with ventricular septal defect, as the case reported. Despite the progress of the treatment, functional limitation survival of these patients remains unsatisfactory. Additional therapeutic strategies are to be targeted to improve the symptoms.

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