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Congenital Heart Disease: Unrepaired Large Patent Ductus Arteriosus Caused With Eisenmenger Syndrome

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Abstract: *The development of Eisenmenger syndrome is due to the unrepaired congenital shunt between the main blood vessels or chambers of the heart. The heart defects that can lead to Eisenmenger syndrome include Atrioventricular canal defect, Atrial septal defect, Patent ductus arteriosus, and Ventricular septal defect. The major complications of Eisenmenger syndrome include cyanosis, polycythemia, arrhythmia, sudden cardiac arrest, heart failure, coughing up blood, stroke, renal problems, endocarditis, and pregnancy complications. A 52-year-old female was admitted with chief complaints of generalized edema and decreased urine output with clubbing of toes over the bilateral feet. The detailed examination of ECHO revealed that she had congenital heart disease (CHD)-large Patent ductus arteriosus (PDA) with bidirectional shunt, severe pulmonary artery hypertension, and right ventricular dysfunction. Treatment was carried out with diuretics, anti-diabetics, sildenafil, thyroxine, dermatological drugs, and neurological drugs.*

INTRODUCTION:

A 52-year-old female presented in the Outpatient Department (OPD) with generalized edema and decreased urine output with clubbing of toes over the bilateral feet.The presence of shunting was confirmed with a decrease in the oxygen saturation (88 %). The patient had a history of Type 2 Cardiorenal syndrome (CRS), Diabetes Mellitus for the past 2 years, and hypothyroidism. She had a Patent Ductus Arteriosus (PDA) that was not repaired because of severe Pulmonary Artery Hypertension (PAH). On physical examination the patient was afebrile and oriented, blood pressure was found to be 100/60 mmHg on both arms and heart rate was found to be elevated (96 bpm) on the day of admission. Normal S1 and S2 with an absence of murmur. Laboratory investigation revealed severe haemolysis and fragmentation of red blood cells (RBC) suggesting the need for blood transfusion. On admission day, her haemoglobin was 10.8 mg/dL and after 3 days it was 10.5 mg/dL. RFT was found to be elevated on all the days (64.2 mg/dl, 71.5 mg/dl, 92.1mg/dl). Laboratory investigation revealed severe haemolysis and fragmentation of red blood cells (RBC) suggesting the need for blood transfusion. The lipid profile recorded indicated HDL at 30 mg/dl and serum cholesterol as 84mg/dl. Dermatology consultation was done for skin lesions and neurology consultation for persistent headaches.

The patient's echocardiography (ECHO) result showed CHD-large Patent ductus arteriosus with shunt reversal, severe pulmonary artery hypertension (PAH), and right ventricular (RV) dysfunction. The inquiry into her past medical history revealed the presence of a large patent ductus arteriosus thus establishing the cause of the Eisenmenger syndrome (ES) in this patient. Hence, all these factors led to the final diagnosis of Congestive Cardiac Failure (CCF).

TREATMENT GIVEN:

Her medication history encompasses Spironolactone and Torsemide combination for heart failure, Sildenafil 20 mg twice daily for pulmonary hypertension, Glimepiride and Metformin combination for diabetes mellitus, Omeprazole for GERD (Gastroesophageal Reflux disease), Thyroxine 12.5mcg o.d. for hypothyroidism, and allopurinol 100mg o.d. for gout. During her hospital days, she was asked to continue her usual own medication except for Spironolactone and Torsemide combination and additionally she was initiated with Furosemide 40 mg intravenously thrice a day (t.i.d) for 6 days to treat Cardiorenal syndrome and for lowering the heart rate Ivabradine 5 mg t.i.d for 6 days. The patient was also given levocetirizine and clobetasol ointment and salicylic acid combination ointment for skin lesions, and the persistent headache was treated with flunarizine. During her hospital days, she was asked to continue her usual own medication except for Spironolactone and Torsemide combination. But during the last two days in the hospital, was given with Torsemide tablet 20mg twice daily and was allowed to continue this along with her past medications.

DISCUSSION:

Eisenmenger syndrome (ES) is an assemblage of symptoms that arise from a congenital heart defect and hemodynamic forces initially result in a left-right shunt, which develops into severe pulmonary arterial hypertension (PAH) and elevated pulmonary vascular resistance (PVR).[1] Due to increased PVR, reversal of the left-to-right shunt takes place and leads to a

right-to-left shunt, thus resulting in significant hypoxemia and cyanosis.[1]In this case, the patient represented the same phenomenon of shunt reversal. Reversal of a left-to-right shunt may result in a bidirectional or right-to-left shunt and thereby subsequently lead to hypoxemia condition which poorly responds to oxygen therapy. The three main reasons behind this reversal are vasoconstriction followed by vascular remodelling and finally thrombosis. The most common defects leading to Eisenmenger syndrome are large atrial septal defects (ASD), large ventricular septal defects (VSD), atrioventricular septal defects (AVSD), and large patent ductus arteriosus (PDA).[1]

The common complaints include swelling, volume retention, syncope, worsening cyanosis, palpitations, or haemoptysis. Chronic hypoxemia can lead to an increase in red blood cell volume and thereby patients may present with symptoms like dizziness, headaches, vision changes, end-stage organ damage, and stroke, indicative in this case where the patient reported persistent headaches.[1] Clubbing in the lower limbs is a hallmark of PDA with reversal of shunt and is often observed in patients.[2] In this case, the patient has clubbing over the bilateral foot. Dermatologic manifestations associated with Eisenmenger's syndrome may include plethora, livedo reticularis, profound acrocyanosis, urate depositions, ecchymosis, and ischemic skin ulcerations[1] which was also noticed in the patient and reported as skin lesions.

Therefore, detailed evaluation of suspected patients should be performed by doing tests like pulse oximetry, chest radiograph, ECG,2D-Colour Doppler ECHO, pulmonary function tests, complete blood count, iron studies, and recording loud murmurs which are typically present in Eisenmenger syndrome.[1] In this situation, the pregnancy should be avoided as it is contraindicated in women who develop PAH with CHD.[3]

Here the patient is diagnosed with Type 2 CRS. It is the chronic Heart failure that results in chronic renal dysfunction. The complication of cardiorenal syndrome include volume overload and the primary treatment option is to removal of fluid either with diuretics or ultrafiltration. High dose of Intravenous(IV) furosemide was effective and safe as it had no adverse impact on renal function.[4]

Cardiopulmonary transplantation is a curative option for Eisenmenger syndrome, but it is impractical to find a donor.[5] Corrective surgery is contraindicated in patients with high PAP and high PVR as it can lead to extensive changes to the pulmonary vasculature.[6] The treatment options include diuretics, antiarrhythmics, anticoagulants, maintaining fluid balance, endocarditis prophylaxis, iron supplements, and trying to avoid precipitating factors like pregnancy, weight gain, high altitude, etc. along with supplemental oxygen and phlebotomy.[1] The usage of supplemental oxygen did not show any mortality benefit, however long-term therapy with Sildenafil has shown improvement in survival of patients with Eisenmenger syndrome.[7]Finally, the patient was discharged with advice to continue medical treatment and to have regular follow-up.

CONCLUSION:

Pulmonary vasodilators like PDE-5 inhibitors are well tolerated and they decrease mortality and improve the haemodynamics and exercise capacity of patients with ES. There are many causes of the disorder and the key is to treat the primary disorder. Once Eisenmenger syndrome develops, the prognosis is poor. Therefore, the initial evaluation of paediatric patients showing both CHD and Pulmonary hypertension should include a comprehensive history and physical examination. With advice to the patient regarding the need for close follow-up.

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