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Case Report

Complementary/Alternative Medicine section

Eisenmenger Syndrome with Unilateral Renal Agenesis: A Rare Case Report

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ABSTRACT

The most advanced form of Pulmonary Atrial Hypertension (PAH), linked with Congenital Heart Abnormalities (CHD), is Eisenmenger Syndrome (ES). This report is about a 26-year-old female who presented with ES treated with Phosphodiesterase type 5 (PDE5) inhibitors, cardiac glycosides, Angiotensin-Converting Enzyme (ACE) inhibitors, loop diuretics, antipyretics, iron and vitamin supplements. Investigations revealed unilateral renal agenesis of left kidney, pulmonary hypertension, ES with large ventricular septal defect and bidirectional shunt. Loop diuretics and PDE5 inhibitors remained the mainstay of the treatment which induced hearing loss and the patient self reported hearing improvement after ceasing furosemide administration. Additional treatment strategy is to be used to improve the symptoms and quality of life of patients.

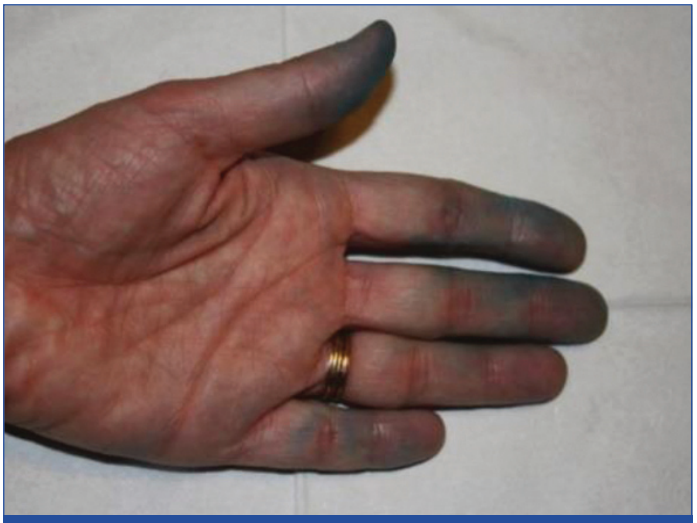
Keywords: Loop diuretics, Pulmonary atrial hypertension, Sildenafil, Ventricular septal defect

CASE REPORT

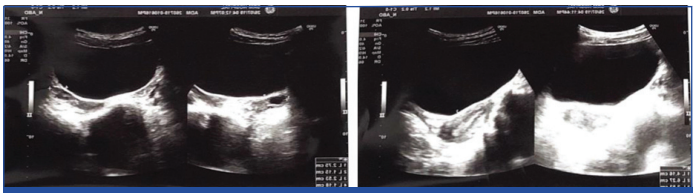
A 26-year-old female patient came to the hospital with the complaints of fever on and off with expectoration, giddiness and cyanosis for four days. The menstrual history revealed four episodes of polymenorrhea every 15 days. The patient is already a known case of ES for past three years and is on regular medication. The patient was prescribed with tab. digoxin 0.25 mg twice a day, tab. Ferrous Sulphate+Folic acid 20/50 mg once a day, tab. ramipril 3.125 mg once a day, tab. sildenafil 5 mg once a day.

On clinical examination, the patient was moderately built and nourished. General examination revealed cyanosis of the hands [Table/Fig-1]. The vitals were stable with a pulse rate of 109/minute and blood pressure was 90/70 mmHg. Her respiratory rate was 24/minute. Breast and abdominal examination revealed no abnormality. Lungs were clear with normal bilateral vesicular breath sounds. Routine investigations revealed erythrocytosis with a haemoglobin of 18.8 g/dL (12-15 g/dL), packed cell volume was 57% (36-46%), platelets were found to be normal 2,25,000 cells/cu.mm (1,50,000-4,00,000 cells/cu.mm), Mean Corpuscular Volume (MCV) 8 fL (83-101 fL), mean corpuscular haemoglobin (MCH) 28pg (27-32 pg), creatinine was 0.6 mg/dL (0.5-1.2 mg/dL), urea was 16 mg/dL (15-40 mg/dL), bleeding time 1 minute 45 seconds and activated clotting time of 5 minutes 2 seconds.The thyroid test revealed an abnormal Thyroid Stimulation Hormone (TSH) of 5.4 micro units/mL.

Echocardiography revealed large ventricular septal defect with bidirectional shunt and moderate pulmonary regurgitation. Electrocardiogram (ECG) revealed sinus tachycardia, right atrial enlargement, and right ventricular hypertrophy. Ultrasonography (USG) of the abdomen showed bicornuate uterus and unilateral renal agenesis of the left kidney [Table/Fig-2]. With the evidence obtained from the investigations, the patient was known to have unilateral renal agenesis, congenital heart disease, ES, large ventricular septal defect with bidirectional shunt and PAH. The patient is currently prescribed with Injection (Inj.). furosemide 20 mg twice a day, tab. digoxin 0.25 mg once a day, tab. carvedilol 3.125 mg twice a day, tab. sildenafil 25 mg once a day, tab. crocin 500 mg twice a day.



[Table/Fig-1]: Cyanosis of hand.



[Table/Fig-2]: USG of pelvis showing bicornuate uterus and unilateral renal agenesis of left kidney.

Differential diagnoses include idiopathic PAH, tetralogy of fallot, tricuspid atresia, transposition of the great arteries, persistent new born PAH, pulmonary infarction and respiratory failure.

DISCUSSION

The Eisenmenger Syndrome (ES) is an intricate multisystem malady which comes about an upshot of a substantial left to right shunt impacting the pulmonary vasculature causing suprasystemic pulmonary artery pressure ensuing a right to left shunt [1]. Nevertheless, patients with ES conventionally get through their third or fourth decades of life, symptoms they may experience includes dyspnoea, cyanosis, clubbing, fatigue, dizziness, and syncope,

curtails their life expectancy. Besides, cardiac arrhythmias, a significant delayed intricacy of ES, are a common cause of unforeseen death in patients with ES. ES does not seem to have prevalence variability neither in males nor females [2]. Prompt prognosis and managing cardiac defect prior to the evolution of PAH is crucial to specific management, if not only supportive care can be held out. The prevalence of ES is about 1-6% in worlds population [3].

The diagnosis of ES is not distinctly convoluted, yet may require cardiac catheterization, an invasive maneuver to compute pressure in the heart and lungs. Further tests comprised of pulse oximetry, which assesses the oxygen levels in blood, chest X-ray, Electrocardiography (ECG), pulmonary function test, iron levels and Complete Blood Count (CBC). Echocardiography principally identifies the heart defect and raises the speculation of elevated pressures in the lungs. Imaging modalities, e.g., cardiac Magnetic Resonance Imaging (MRI) furnishes beneficial anatomic information [4]. The most advanced form of PAH linked with CHD is ES [5]. Cardiopulmonary transplantation is curative for ES which is impractical in most settings.

Pharmacology has ameliorated manifestations, yet not mortality. The pharmacological management often involved the use of cardiac glycosides, diuretics, anti-arrhythmic agents and/or anticoagulants; hardly these approaches profoundly modified survival or risk of debilitation. Cardiopulmonary transplantation is curative for ES which is impractical in most settings. Warfarin has been traditionally availed for anticoagulation in Eisenmenger syndrome. Surgical rectification of the causative cardiac deformity in adult patients is principally inappropriate. Pharmacology has ameliorated manifestations, yet not mortality. Warfarin has been traditionally availed for anticoagulation in ES. Surgical rectification of the causative cardiac deformity in adult patients is principally inappropriate [6].

Phlebotomy with isovolumic replacement is utilised in the management of hyperviscosity syndrome interconnected with the elevated production of red blood cells; however, only patients with specific signs of hyperviscosity should be phlebotomized [7]. Co-administration of sildenafil with furosemide may induce hearing loss which is of sensorineural origin by Cyclic Guanosine Monophosphate (cGMP) elevation mediated cochlear toxicity. The index patient self reported hearing improvement after discontinuing sildenafil, which proves that this hearing loss is reversible. Hence, careful discretion and monitoring is advised if at all concomitant administration of ototoxic agents such as loop diuretics/CYP3A4 inhibitors, and PDE5 inhibitors is mandated.

Clinical follow-up must spotlight on investigating yearly CBC, iron examinations, renal function, and uric acid, alongside focuses on revising any anomalies. Patients should also be evaluated with pulse oximetry, both with and without supplemental oxygen. Any aberrations recommending hypoxemia mandates another estimation.

CONCLUSION(S)

Early diagnosis and surgical treatment for ES bring about a favourable prognosis, whereas, a late diagnosis with the development of pulmonary hypertension and heart failure has a negative prognosis. Heart failure, cardiac arrhythmia, and thromboembolic cerebrovascular illness kill the majority of patients. Drugs like sildenafil and furosemide can help patients with ventricular septal defect improve their prognosis and quality of life. Despite progress in treatment, these patient's functional limitation survival remains dismal. To improve the symptoms, more therapy measures to be pursued.

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