

A Rare Case of Eisenmenger Syndrome in a Young Adult Diagnosed with Dengue Fever

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Abstract: Eisenmenger syndrome¹ is a constellation of symptoms that arise from congenital heart defects. It occurs when a long standing left to right anatomic cardiac shunt caused by congenital heart defect (typically VSD and ASD and less likely PDA) causes pulmonary hypertension and eventually reversal of shunt into a cyanotic right to left shunt, manifesting in early life. This case discusses the clinical presentation, diagnostic workup, and management of a patient of Eisenmenger syndrome secondary to an uncorrected ventricular septal defect (VSD) that presented with Acute Dengue Fever. A 22 year old female presented to the emergency department with history of self induced abortion at home and chief complaints of fever with chills, dyspnea on exertion NYHA Grade 3, giddiness and ghabraman. Evaluation of this patient revealed hypoxemia, central cyanosis, clubbing, cardiomegaly and VSD with a bi-directional shunt and moderate PAH. Patient was given oxygen support (initially for stabilization), medical care², blood products, eventually anticoagulation, and finally, referred for surgical care³.

Keywords: Eisenmenger syndrome, Congenital Heart Defect, Dengue fever, Cyanosis

1. Introduction

Eisenmenger syndrome¹ is a constellation of symptoms that arise from congenital heart defects. It occurs when a long standing left to right anatomic cardiac shunt caused by congenital heart defect (typically VSD and ASD and less likely PDA) causes pulmonary hypertension and eventually reversal of shunt into a cyanotic right to left shunt, manifesting in early life. This case discusses the clinical presentation, diagnostic workup, and management of a 22 year-old female with Eisenmenger syndrome secondary to an uncorrected ventricular septal defect (VSD) presented with Acute Dengue Fever.

2. Case Report

A 22 year old female presented to the emergency department with history of self induced abortion at home and chief complaints of fever with chills, dyspnea on exertion NYHA Grade 3, giddiness and ghabraman since 3 days. There was no history of cough, expectoration, hemoptysis, chest pain, pedal oedema, blood transfusion, substance or toxin exposure. There was no other significant medical or personal history.

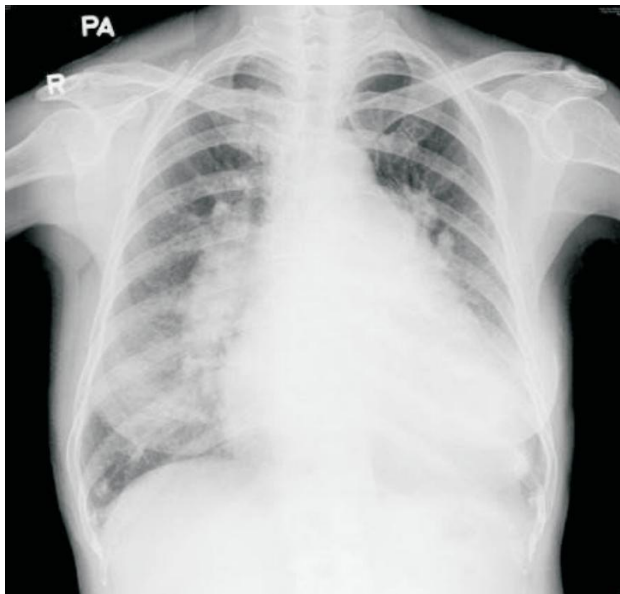
On examination, the patient was conscious and oriented to time, place and person with normal temperature, tachypneic with respiratory rate of 24/minute, Oxygen saturation of 71% on room air, a regular pulse of 64/minute, and a blood pressure of 104/74 mm of Hg. Inspection revealed grade 4 clubbing and central cyanosis with bluish discoloration of lips, checks and oral mucosa. Upon auscultation, S1S2+ with a loud P2 component, respiratory system auscultation was clear with bilateral air entry.



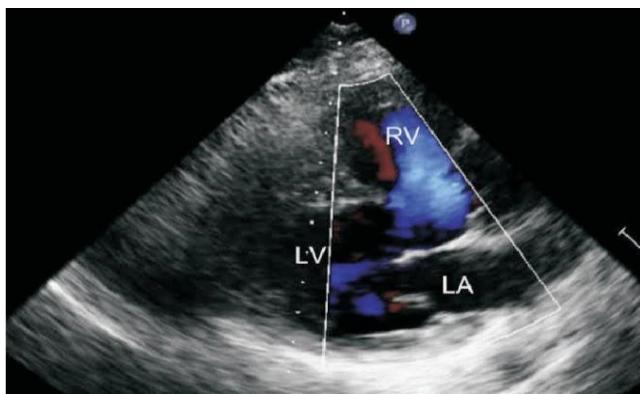
| Blood investigations | |
|-------------------------------|----------|
| Hemoglobin (mg/dl) | 7 |
| Platelets (/mm ³) | 51,000 |
| WBC(/mm ³) | 6380 |
| Creatinine (mg/dl) | 0.86 |
| SGPT(IU/dl) | 38 |
| Dengue Igm | Positive |
| pH | 7.35 |
| SpO2(%) | 72.7 |
| pO2(mm of Hg) | 41 |

ECG showed ST -T changes in V1-V6 leads.

Radiological findings were chest X-ray suggestive of cardiomegaly. Abdominal-pelvic ultrasound showed retained product of conception.



2D echocardiography- Large VSD with Bidirectional shunt, Moderate TR, Moderate PAH RSV: 70mmHg, RA, RV dilated.



Management²:

In treating this patient, Initially, she was kept on Bipap support due to low O₂ saturation. However the patient was weaned off O₂ support and maintained saturation of 70-72% on room air. Patient was treated with guarded fluids and antipyretics with strict temperature charting for dengue fever. One unit of PCV and 4 units of PRC were given in view of low hemoglobin and falling platelet count respectively. Following which misoprostol was given and dilation & evacuation for retained product of conception. As the disease causes a hypercoagulable state, anticoagulation with warfarin was given. Once the patient had become stable, she was further referred for surgical management of VSD³.

3. Discussion

The most common defects leading to Eisenmenger syndrome are ASD, VSD, and PDA defects. The following three main processes result in the ultimate reversal of a left-to-right into a right-to-left shunt. Eisenmenger syndrome is a relatively rare disorder that is usually seen in persons with poor healthcare access (i.e., rural areas), in whom large anatomical defects may go undetected for many years Initially the congenital heart defects lead to left to right shunting of blood

causing increased pulmonary blood flow (endothelial dysfunction and smooth muscle proliferation). There is marked rise in peripheral vascular resistance and hence results reversal of shunt i.e left to right shunt manifesting Eisenmenger syndrome.

4. Conclusion and Implications

Due to similar presentation and resemblances of the underlying pathophysiology, other causes of pulmonary hypertension should be ruled out. Certain rheumatological and autoimmune diseases, such as mixed connective tissue disorder, scleroderma, and systemic lupus erythematosus, may have similar presentations and hence, are important differentials. Hepatitis B, C, and HIV serologies also must be considered, as these may have systemic vascular presentations.

In patients with Eisenmenger syndrome, certain conditions should be avoided such as pregnancy, dehydration, isometric exercise, iron deficiency anemia, and significant time spent at high altitudes. When prescribing antihypertensives, caution should be exercised when using peripheral vasodilating agents, which may cause worsening of the right-left shunt.

References

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