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Case Report

Sickle Cell Crisis presenting as Acute Reversible Pulmonary Arterial Hypertension

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Abstract

Pulmonary arterial hypertension (PAH) is a relatively frequent and severe complication of sickle cell (SC) disease and an independent risk factor for mortality. We report case of SC trait presented with acute onset dyspnea class IV, hypoxia, and moderate PAH with dilated right atrium and right ventricle on 2DECHO. Normal computed tomography Pumonary angiography on 2DECHO and normal CT pulmonary angiography. He was treated with hydration, O_2, hydroxyurea, and folic acid. The patient improved symptomatically and 2DECHO was normalized.

Key words: Hydration, Reversible pulmonary arterial hypertension, Sickle cell crisis

INTRODUCTION

Sickle cell disease (SCD) encompasses a group of hemoglobinopathies characterized by amino acid substitutions in the beta-globin chain. The most frequently occurring form of SCD is caused by homozygous presence of hemoglobin S (HbSS). Pulmonary hypertension (PH) is a relatively frequent and severe complication of SCD and an independent risk factor for mortality.[1]

Prevalence

Echocardiographic screening studies have identified evidence of elevated pulmonary pressures, defined as a tricuspid regurgitant jet velocity ≥2.5 m/s (equivalent to a pulmonary artery systolic pressure of approximately 36 mmHg), in 30–40% of HbSS and 10–28% of HbSC adults.[2]

Pathogenesis

The exact pathogenesis of Pulmonary arterial hypertension (PAH) in SCD is not known, but a number of potential contributing factors have been implicated, including endothelial injury from recurrent sickling, acute and chronic inflammation, hypercoagulability, chronic intravascular hemolysis, and altered bioavailability of the potent vasodilator nitric oxide (NO). Vascular remodeling caused by chronically elevated left heart pressures from diastolic dysfunction may also contribute, similar to PH group 2, which is purely due to left heart disease.[3]

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CASE REPORT

A 40-year-old male known case of Sickle cell trait was on hydroxyurea, folic acid, and tablet sodamint, which was presented with acute onset dyspnea of 2 days duration without fever, cough, orthopnea, leg edema, and angina. Physical examination revealed tachycardia with pulse rate of 130/min, regular and good volume, blood pressure was 100/70 mmHg, no cyanosis, clubbing, chest was clear, CVS-S1, S2 was normal and no murmur, Spo_2 was 88% at room air, ECG showed sinus tachycardia with T wave inversion in anterior leads [Image 1], and 2DECHO [Image 2] showed dilated right atrium/right ventricle (RA/RV) with moderate PAH (65 mmHg). Acute pulmonary thromboembolism (PTE) was suspected, so computed tomography pulmonary angiography (CTPA) was done. To surprise, it was normal. Hence, the patient was treated with hydration, O_2, low molecular weight heparins,

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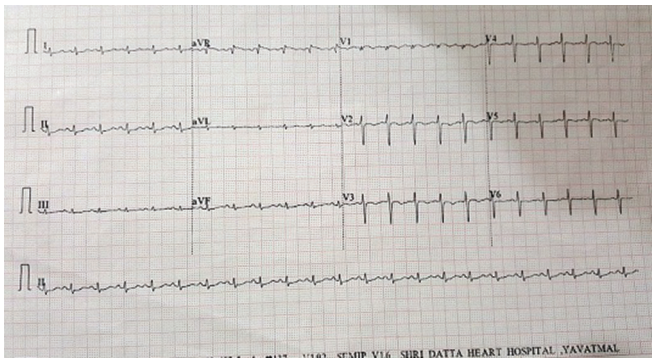

Image 1: ECG showing sinus tachycardia and RV strain

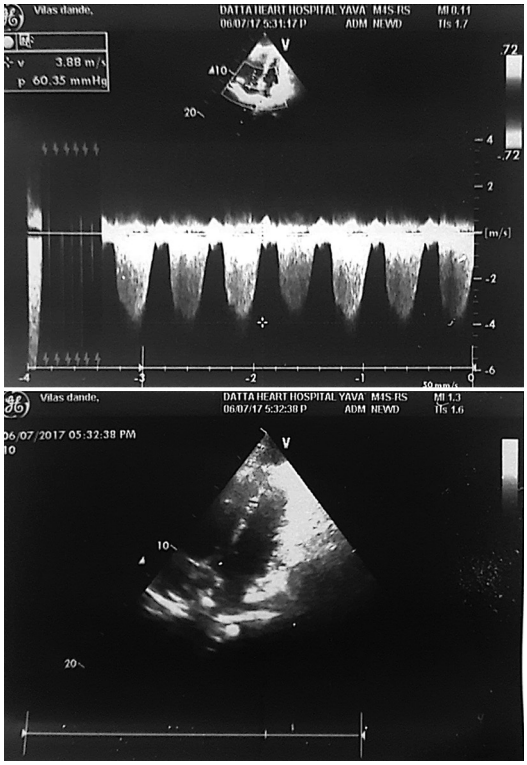

Image 2: Dilated RA and RV with moderate PAH

hydroxyurea, and supportive drugs. The patient improved symptomatically, O_2 was stopped, and repeat 2DECHO [Image 3] showed normalization of dilated RA/RV and pulmonary arterial pressure (PAP).

We conclude that this normalization of PAP was due to normalization of PVR which might have increased due to occlusion of capillaries by deformed red blood cells.

DISCUSSION

PAH is defined as an elevated mean arterial pressure ≥25 mmHg at rest.[1,4] PAH has several etiologies and can be a progressive, fatal disease, if untreated.

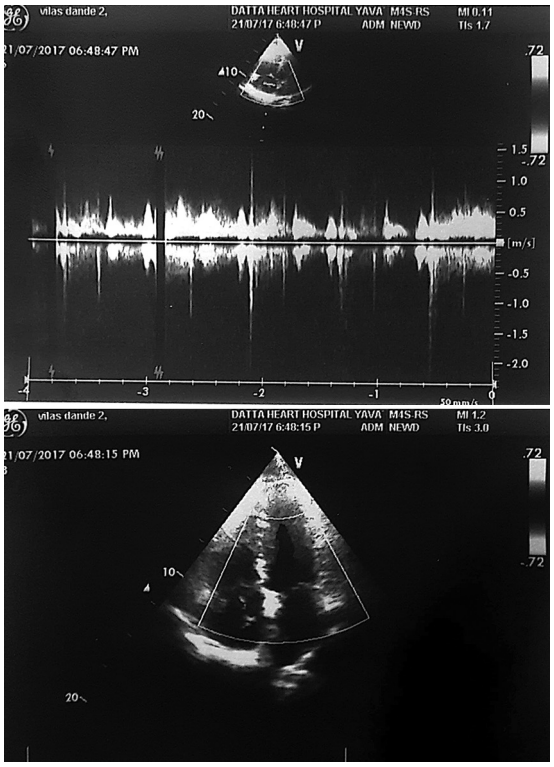

Image 3: Follow up ECHO showing normalisation of RA, RV and PA pressure

The cause of PH, which has been reported in 20–32% of SCD patients, is multifactorial, with contributing factors including hemolysis, impaired NO bioavailability, chronic hypoxemia, high cardiac output, thromboembolism, and parenchymal and vascular injury caused by sequestration of sickle erythrocytes, chronic liver disease, and asplenia.[3] There are very few instances, where PAH is reversible like acute PTE.

We report a case, where the patient presented with acute dyspnea and moderate PAH. The clinical presentation was like acute PTE, but CTPA was did not show any thrombus. PAH regressed to normal, 2DECHO picture of dilated RA and RV normalized with hydration and oxygenation. Transesophageal echo showed intact IAS and no evidence of sinus venosus ASD. This patient was known case of sickle cell (SC) trait, so we conclude that acute reversible PAH was secondary to SC crisis. We have searched in the literature for PAH in SCD, it showed chronic PAH but have not seen a case which presented like this.

There is a one case series of six patients by Abraham *et al.* which mention about regression of PAH by oxygen supplementation in Chronic Bronchitis. In my case also prolong, oxygenation had helped in PAH regression.[5]

Intensification of hydroxyurea and exchange transfusion is mainstay of treatment, we have insufficient data to explain, whether prolonged oxygenation can be a therapy for chronic PAH.

CONCLUSION

Acute reversible PAH is seen in acute PTE which can improve with early thrombus resolution. Similar finding are observed in sickle cell crisis. Early treatment of crisis can help to normalize PAP and to prevent development of chronic PAH. In our observation, this is a second cause of acute reversible PAH after acute PTE.

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