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A Case Report

An unusual presentation of chronic eosinophilic leukemia as pulmonary arterial hypertension- A Case Report

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ABSTRACT

***P**ulmonary arterial hypertension (PAH) as a manifestation of chronic eosinophilic leukemia is a rare presentation. We present a case of thirty-four years old male who was diagnosed with chronic eosinophilic leukemia manifesting as severe pulmonary artery hypertension and had a dramatic response with imatinib therapy. Chronic eosinophilic leukemia may rarely manifest as pulmonary arterial hypertension and can have clinical and haematological remission with Imanitib therapy.*

Keywords: *Pulmonary artery hypertension; chronic eosinophilic leukemia; Imatinib; Echocardiography*

1. INTRODUCTION:

Pulmonary arterial hypertension (PAH) as a spectrum of chronic eosinophilic leukemia is a rare presentation has only been suggested by case reports and few case series with exact incidence and prevalence of PAH in such population is yet to be defined.1 Chronic myeloproliferative diseases (CMPD) associated with PAH is now included in the group 5 category, corresponding to PAH for which the etiology is unclear and/or multifactorial.2 Identifying the underlying disorder and targeting the PDGFR with imatinib can be a promising therapeutic armamentarium in treating PAH secondary to myeloproliferative disorders.3 We describe one such unusual presentation of chronic eosinophilic leukemia with PAH who had significant resolution of his symptoms, improvement in his functional class, six-minute walk distance and Pulmonary artery (PA) pressures along with normalization of his haematological parameters after 4 weeks of imatinib therapy.

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2. CASE REPORT

Thirty years old gentleman with no known risk factors for cardiovascular or pulmonary disease presented with a history of insidious onset, gradually progressive, shortness of breath (WHO Class III) along with fatigue for last 1 month. He had no history of chest pain, presyncope/syncope or palpitations. The patient denied any history of fever, joint pains, abnormal discolouration of urine, and prolonged intake of any drug or recent travel. On examination, BP= 130/80 mm Hg, PR=90/min, regular, with all peripheral pulses palpable. There was no pallor, icterus, clubbing, cyanosis or peripheral edema. Jugular venous pressure was not raised. Cardiovascular examination revealed loud second heart sound with grade III pansystolic murmur heard at the right lower parasternal area, no other murmur was heard, no S3/S4, pericardial rub or added sounds. ECG (Figure 1) revealed sinus rhythm, right axis deviation, and was consistent with right ventricular hypertrophy.

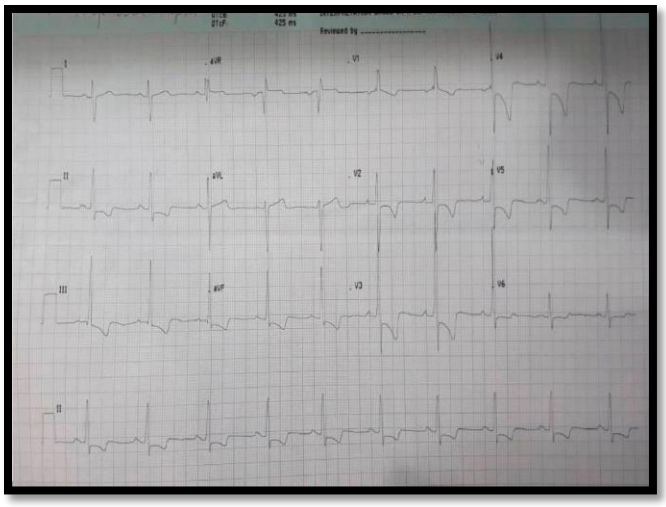

Fig 1: ECG of the patient showing normal sinus rhythm with right axis deviation and right ventricular hypertrophy with strain.

Chest X-ray (Figure 2) had evidence of pulmonary artery dilation with RA enlargement with no evidence of pulmonary venous hypertension (PVH) or other signs of lung disease. Echocardiography showed Normal LV dimensions, No Regional wall motion abnormalities with left ventricular ejection fraction (LVEF) of 58 %. Flattening of the interventricular septum was noted with enlarged main pulmonary artery (MPA), MPA diameter in parasternal short-axis view was measured to be 39 mm. There was severe tricuspid regurgitation with Peak tricuspid velocity of 4.5 m/sec and RVSP of 94 mm Hg and pulmonary artery end-diastolic pressure (PAEDP) of 30 mm Hg and right ventricular acceleration time was 102 msec. IVC diameter was 22 mm with less than 50 % collapse on inspiration. CECT Thorax (Figure 3) showed right atrium (RA), right ventricle (RV) and pulmonary artery (PA) dilatation, (PA diameter 41 mm, PA / Ascending aorta diameter ratio was 1.57) with no evidence of thrombus. There was no abnormal lung parenchyma, pleural effusion, hemangioma or any other abnormality. Complete blood count analysis revealed haemoglobin of 16.3 g/dl, total white blood cell count of 8,300 per microlitre with 37 % polymorphs, 18 % lymphocytes, 37 % eosinophils, 6 % monocytes and 1 % basophils. Peripheral smear showed marked eosinophilia with thrombocytopenia with an absolute eosinophil count of 3,070 per microlitre and platelet count of 65,000 per microlitre of blood with the absence of any microfilaria or any other parasite. Serum vitamin B12 was 1463 pg/ml which was elevated. Serum folate levels, thyroid function, liver function, renal function tests were normal. Stool examination was unremarkable for any ova, cysts or parasites. The patient underwent bone marrow aspiration examination which revealed cellular marrow with trilineage hematopoiesis and marked eosinophilia. FIP1LPDGFRA was negative with bone marrow findings consistent with chronic eosinophilic leukemia. The patient was started on intravenous methylprednisolone therapy for three days and imatinib 100 mg per oral once daily. The patient had significant relief in his symptoms with improvement in 6-minute walk distance and functional class, normalisation of platelet count and reduction in absolute eosinophil count. His echocardiography after 4 weeks of therapy showed peak tricuspid regurgitation velocity of 3.4m/sec and RVSP of 56 mm Hg. No significant side effects to therapy were reported by the patient.

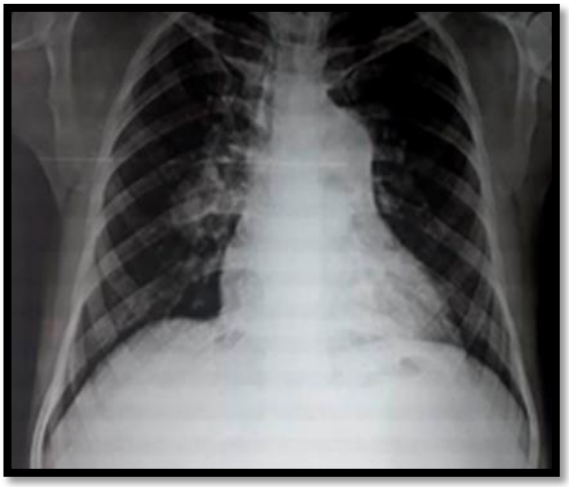

Fig 2: Chest X Ray (PA View) shows pulmonary artery dilation with RA enlargement with no evidence of PVH or other signs of lung disease.

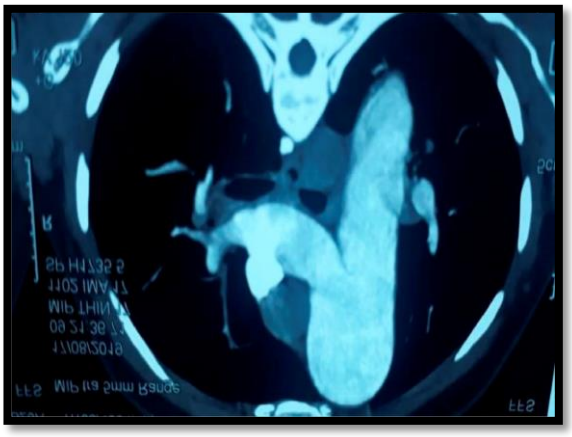

Fig 3: Computed tomography thorax showed right atrial, right ventricular and pulmonary arterial dilatation (PA diameter 41 mm, PA / Ascending aorta diameter ratio was 1.57) with no evidence of thrombus.

3. DISCUSSION

The clinical classification of pulmonary hypertension comprises of various clinical conditions which often have similar clinical features, pathophysiology and management. Chronic myeloproliferative diseases (CMPD) associated with PAH is currently classified in group 5 in which the aetiology is unclear and/or multifactorial.4 However, the possible association of PAH with CMPD has only been suggested by case

reports and small case series.5-6 Our patient was diagnosed to have PAH with a possible association with chronic eosinophilic leukemia and responded well to imatinib therapy on follow up with an improvement in symptomatic profile, laboratory and echocardiographic parameters. Several causal factors may be implicated in the pathogenesis of PAH in such patients. Increased expression of platelet-derived growth factor receptor (PGDFR) leading to smooth muscle hyperplasia is one of the several pathophysiological pathways that have been proposed.1 Imatinib, a potent inhibitor of tyrosine kinases including BCR-ABL and PDGFR7, therapy directed for haematological remission, also resulted in alleviation of symptoms due to PAH with inhibition of PGDFR as one of the plausible mechanisms. Thus, targeting the PDGFR with imatinib is a promising therapeutic armamentarium in treating PAH secondary to myeloproliferative disorders.

4. CONCLUSION

We describe one such unusual presentation of chronic eosinophilic leukemia with PAH who had significant resolution of his symptoms, improvement in his functional class, six-minute walk distance and Pulmonary artery (PA) pressures along with normalization of his haematological parameters after 4 weeks of imatinib therapy.

5. FINANCIAL DISCLOSURE

None.

6. DECLARATION OF CONFLICT OF INTEREST

None.

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