

A CASE OF POLYCYTHEMIA VERA

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INTRODUCTION

Polycythemia vera (PV) is a clonal disorder causing overproduction of all three-haematopoietic cell lines, most prominently the red cells in the absence of a recognizable physiological stimulus.

Erythropoiesis, megakaryocytopoiesis and granulocytopoiesis are all increased. PV usually presents with non-specific complaints like headache, tiredness, vertigo, visual disturbances, epigastric burning or thrombotic phenomena. Pruritis is common especially after a warm bath [2].

There are no population-based studies on PV in our country. Data from AFIP during April 2005 to April 2006 showed a total of 48 cases of erythrocytosis, out of which 14 were found to have PV. There was slight male predominance and median age was 58 years.

We report this case because we want to emphasize the importance of early diagnosis and adequately selected treatment as it greatly affects the prognosis.

CASE REPORT

A 65-year-old house wife resident of a village near Attock, presented to Military Hospital, Rawalpindi in Nov, 2006 with bruises on both arms for ten days and epigastric burning for two months. The epigastric burning was aggravated by spicy food and associated with excessive belching. There was no history of bleed from any other site except easy bruising. Nine months earlier she presented to a local hospital with history of pain and swelling right leg of three days duration. Suspecting deep venous thrombosis, her Doppler ultrasound (USG) of right leg was done which showed thrombus in right femoral vein and decreased flow in right femoral artery. She was advised anticoagulation therapy for six months but

she discontinued it after four months with apparent complete recovery. She was a known hypertensive for ten years, taking irregular medication. She did not smoke. There was no history of haematological disorders in the family. On examination, her pulse was 76 beats per minute, regular; blood pressure 130/80 mm of Hg; respiratory rate 16/min and she was afebrile. She had a plethoric look (Fig. 1), conjunctival injection; ecchymoses on both arms (Fig. 2) and her right dorsalis pedis artery and posterior tibial arteries were not palpable. Fundoscopy revealed bilateral hyperemic discs with engorgement of retinal vessels.

CBC showed Hb 16.9g/dl, erythrocytosis (TRBC $9.4 \times 10^9 /L$), leucocytosis (TLC $15.8 \times 10^9 /L$), thrombocytosis (platelets $475 \times 10^9 /L$) and elevated haematocrit (59%) alongwith hypochromia and microcytosis. PT (13/13), PTTK (34/34), INR (1.0), LFTs, serum urea, creatinine, electrolytes, blood glucose fasting, serum lipid profile, urine RE and chest x-ray were within normal limits. ECG showed partial left bundle branch block (LBBB) with occasional VPCs. Echocardiography revealed left ventricular hypertrophy with good left ventricular function. Pulse oximetry showed O₂ saturation of 99% at room air. Tc-99 m labeled red cell mass showed a value of 66.32 ml/kg body weight (normal 30+5 ml/kg body weight). Serum erythropoietin (EPO) level was reduced to 2.8 mIU/ml (ref range 3.3-16.6mIU/ml). USG abdomen showed splenomegaly (spleen size 12.6cm) and a small right kidney measuring 4.7cm while left kidney was normal in size. The small right kidney was found to be congenitally hypoplastic as no vascular supply was detected on renal doppler USG. DTPA renal scan showed non functioning right kidney and well functioning left kidney with

compensatory hypertrophy. On Doppler USG, right common iliac artery was thrombosed soon after its origin but the collaterals were well developed. Angiography confirmed the blockade of right common iliac artery at its origin but the limb was well perfused through collaterals. Haemoglobin electrophoresis did not reveal any disorder. Bone marrow examination showed trilineage hyperplasia with erythroid prominence, platelet clusters and absent iron stores suggestive of myeloproliferative disorder – Polycythemia Vera (Fig. 3). LAP score was 256 (ref range < 300), uric acid was 483 μ mol / L, vitamin B12 was 586 pg / ml. Serum Ferritin (20 ng /ml) and Iron (9.2 μ mol / L) were on the lower normal limit, while serum TIBC (52 μ mol/ L) was on the upper side. Upper GI endoscopy revealed mild antral gastritis and histopathology of the biopsy taken showed

chronic nonspecific gastritis. The patient, hence, fulfilled the PV Study Group Diagnostic Criteria.

The Patient was managed with serial phlebotomies, capsule hydroxyurea 500 mg daily, low dose aspirin 75 mg daily, allopurinol 300 mg daily, omeperazole 20 mg twice daily and amlodipine 10 mg daily.

The patient was followed up for 03 months. There was symptomatic improvement, plethora decreased (Fig. 1) and blood counts improved. (Hb 14.9 g/dl, TLC $12.5 \times 10^9/L$, platelets $341 \times 10^9/L$ and HCT 50%).

DISCUSSION

Most patients of PV in the early disease present with symptoms due to hyperviscosity like plethora, headache and visual disturbances. Thrombotic phenomena are a frequent presentation and are more common

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After



Fig.1a: Plethora before after treatment.

Fig. 1b: After treatment



Fig. 2: Ecchymosis on both arms

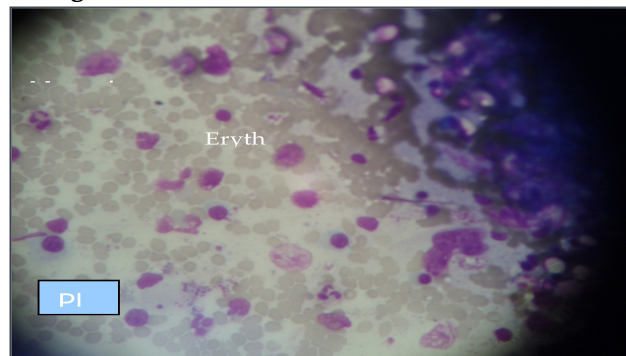


Fig. 3: Bone marrow biopsy suggestive of polycythemia vera

in women. Paradoxically, polycythemia vera patients also have haemorrhagic tendency but haemorrhagic manifestations are far less common. Spleen is palpable in 70% of cases but nearly always enlarged when imaged [2].

Polycythemia is suspected if Hb is more than 16.5 g/dl in females and 18.5 g/dl in males with HCT >48% and 52% respectively [2]. RBC mass should be done in all unless HCT > 60 % [3]. EPO level may be normal or reduced. Low levels have specificity of 92-99% [4]. Oxygen saturation is normal. Bone marrow examination becomes more important if there is a change in clinical course of the disease. It distinguishes PV from secondary polycythemia [5]. Serial phlebotomies are the mainstay of treatment, with aim to reduce HCT to <45% in men, <42% in women and <36% in pregnancy [6]. Hydroxyurea is used as an adjunct to phlebotomy especially in patients prone to thrombosis like our patient [7]. Interferon is reserved for high-risk women of child bearing potential and for refractory pruritis [8]. Alkylating agents and radioactive phosphorus ³²P should be considered only in elderly, as they are highly leukemogenic [9]. Low dose aspirin (75-100 mg/day) should be given to all if not otherwise contraindicated [10, 11]. Our patient benefited from serial phlebotomies, hydroxyurea and low dose aspirin.

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