www.jmscr.igmpublication.org

Impact Factor 3.79
Index Copernicus Value: 5.88
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: http://dx.doi.org/10.18535/jmscr/v3i12.11

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

# Polycythemia Vera Presenting As Acute Ischemic Stroke: A Case Report

Authors
**Dr Siba Prasad Dalai[1], Dr Lalit Kumar Meher[2], Dr Sameer Panda[3], Dr Pankaj Kumar Hui[4], Dr Sujit Kumar Tripathy[5]**
[1]PG Trainee, Dept. of Medicine, [2]Professor & HOD PG Dept. of Medicine
[3]PG Trainee, Dept. of Medicine, [4]Associate Professor, Dept. of Medicine
[5]Senior Resident, Dept. of Medicine
[1,2,3,4,5]Department of General Medicine, MKCG Medical College and Hospital
Corresponding Author
**Dr Siba Prasad Dalai**
PG Trainee, Department of Medicine, MKCG Medical College, PG Hostel 2, Room No. 18, MKCG Medical College
Contact No. – 8895182567, Email Id: *drsibadalai@gmail.com*

**Abstract**
*Polycythemia Vera (PV) patients are prone for vaso-occlusive events including cerebral ischemia. Although infrequent, Polycythemia Vera can present as a case of acute stroke for the first time. A 54 year old male presented to us with left sided hemiparesis and imaging revealed lacunar infarct in right middle cerebral artery (MCA) territory. Evaluation showed it as a case of Polcythemia Vera. The case is reported in view of rarity of stroke being the inaugural manifestation of Polycythemia Vera.*
**Key Words**: *Polycythemia Vera, Ischaemic stroke, Erythropoetin*

## INTRODUCTION

Stroke is the third leading cause of mortality and morbidity worldwide. Ischemic stroke may be the first presenting symptom of Polycythemia Vera in 15% or more of those affected.(1) Here we report a case of Polycythemia Vera presenting with acute ischemic stroke as the initial symptom.

## CASE REPORT

A 54 year male non-diabetic, normotensive, presented to us with sudden onset weakness of left upper and lower limbs and slurring of speech for 2 days. There was no history of head trauma, seizure, lossof consciousness or similar episodes in the past. There was no history of fever, chest pain or tightness, skin rash, bleeding manifestations, blurring vision, leg swelling, itching and significant headache. He was a non-smoker. On examination, he was conscious and oriented, pulse was 76/min, regular. Blood pressure was 130/84 mm of Hg in right arm supine position. On examination he had congested conjunctiva and redness of hand. (Fig.1). There was no pedal edema, cyanosis, clubbing, lymphadenopathy or any other significant abnormality. Neurological examination revealed left sided upper motor type facial palsy, power on left upper and lower limbs was 3/5 with brisk deep

tendon jerks and left plantar reflex was extensor. On the right, upper and lower limbs power was 5/5 with normal deep tendon reflexes and plantar response was flexor. There was mild dysarthria. Chest and cardiovascular and abdominal system examination revealed no abnormality other than moderate splenomegaly.

With the provisional diagnosis of CVA with left sided hemiparesis, relevant investigation were planned. Non Contrast CT scan of brain showed lacunar infarct in right MCA territory. (Fig. 2)

Complete blood count (CBC) study revealed Hb = 20.3 gm%, TRBC = 10.02 x 106 /µL, TWBC= 15.96 × 103 / µL with neutrophil 70%, TPC = 210 ×103 /µL, Hematocrit = 61.9 % , MCV = 61.8 fl, RDW-SD = 57.7 Fl (Ref range 37-54) and RDW-CV =27.1 % (Ref range 11.5-14). Peripheral blood film showed neutrophilic leukocytosis without any premature cells. ESR was 02 mm in 1st hour. Serum uric acid was 7.19 mg/dl. Bone Marrow Aspiration study showed panmyelopoesis. Serum erythropoietin level (by CLIA method) was 3.90 mIu/mL. (Ref range 4.3-29). Fasting plasma sugar,2hour PPBS, fasting lipid profile, blood urea, serum creatinine and serum electrolytes were within normal range. ICT for malaria, HBsAg, Anti HCV, HIV were negative. USG of Abdomen and pelvis showed moderate splenomegaly without any focal lesion. X-Ray of chest, ECG and 2D Echocardiography were normal. An assay for JAK2 V617F came out to be positive.

With elevated RBC count, PCV, Hb%, leukocytosis, microcytic erythrocytosis with increased RDW, BM Aspiration study suggestive of panmyelopoesis, reduced serum erythropoietin level and absence of hypoxia and other apparent secondary cause of erythrocytosis, the case was diagnosed as Polycythmia Vera in accordance with revised WHO Criteria. Our patient beingnon-obese, normotensive and non-diabetic without dyslipidemia and no other significant risk factors for athero-thrombosis and no evidence of embolism in echocardiography, Polycythemia Vera was ascertained to be the underlying cause of ischemic stroke. The patient was put on low dose aspirin and venesection was performed.

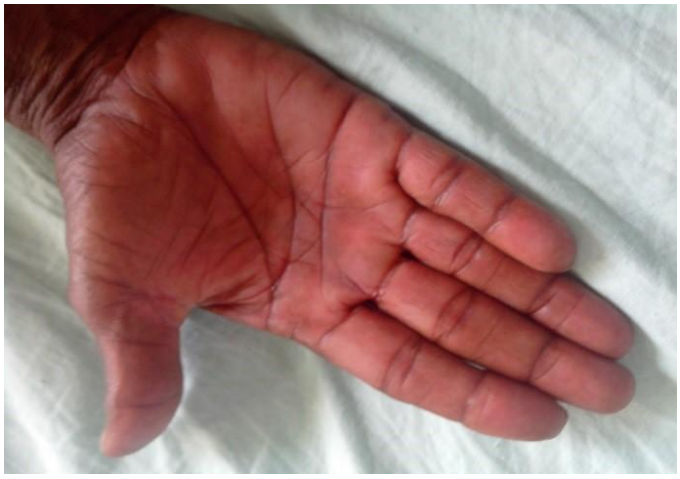

**Fig 1.** Ruddy cyanosis of palm

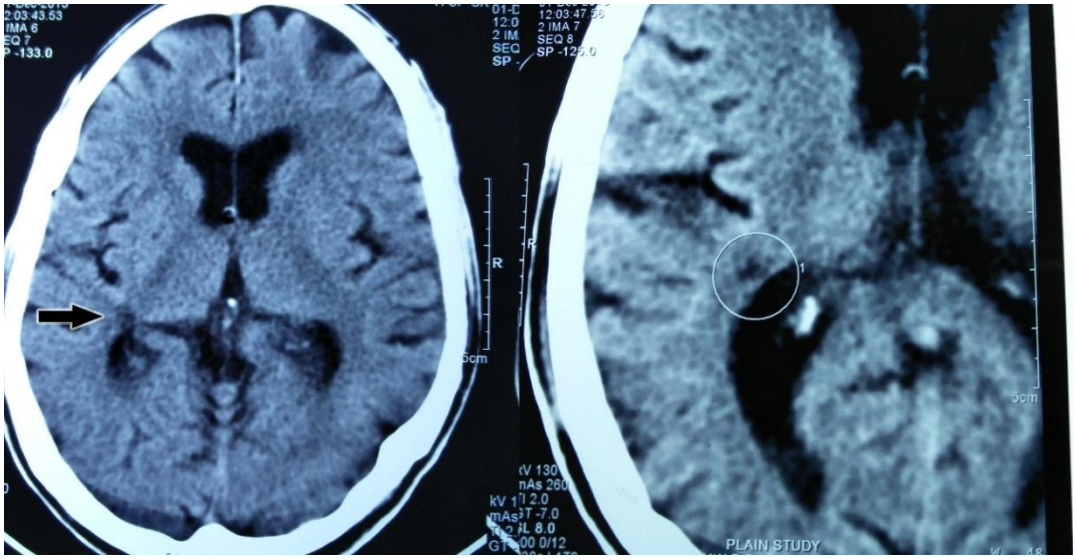


**Fig 2.** NCCT scan brain showing lacunar infarct in right MCA territory

## DISCUSSION

Polycythemia Vera is a clonal disorder involving a multipotent hematopoietic progenitor cell in which phenotypically normal red cells, granulocytes and platelets accumulate in absence of a recognizable physiologic stimulus. The patients of PV commonly present with nonspecific symptoms like fatigue, headache, dizziness, vertigo, tinnitus, pruritus or blurring of vision. Thrombotic phenomena are a frequent presentation including cerebral arterial and venous thrombosis causing stroke. The most likely mechanisms are hyperviscosity and impaired cerebral blood flow. An inverse relationship between cerebral blood flow and the packed cell volume has been shown in Polycythemia Vera. Cerebral blood flow significantly reduces impairing normal uptake of oxygen by tissues and thus impairing microcirculation.[(2)] Moreover, an increased hematocrit leads to decreased reperfusion and increased infarct size.

Treatment of ischemic stroke with PV should aim at reducing further clot formation by reducing high blood viscosity[(3)]. Serial phlebotomies are the mainstay of treatment, with aim to reduce hematocrit to <45% in men, <42% in women and <36% in pregnancy. Hydroxyurea is used along with phlebotomy in patients prone to thrombosis. Pegylated Interferon alpha produces complete responses in PV patients and is recommended for use in the presence of splenomegaly, thrombocytosis, pruritus and thrombo-hemorrhagic complications.[(4)]Imatinibmesylate decreases the frequency of requirement of phlebotomy as well as decreases the spleen size.[(4)] Low dose aspirin (75-100mg/day) should be given to all patients if not otherwise contraindicated[(5)]. Anagrelide, a phosphodiesterase inhibitor reduces the platelet count and is protective against venous thrombosis. Histone Deacetylase inhibitor, Givinostat down regulates JAK2V617F protein. Other treatment includes Busulfan and Ruxolitinib. Splenectomy can be done in patients with massive splenomegaly unresponsive to medical therapy. Allogenic bone marrow transplantation may provide a cure in young patients.

Only a handful of case reports are present depicting acute ischemic stroke as the initial presentation of PV.[(6,7)] In conclusion, PV and other myeloproliferative disorders must be considered while evaluating any case of acute ischemic stroke and treated accordingly.

**CONFLICTS OF INTEREST**: NIL

## REFERENCES


1. Gruppo Italiano Studio Policitemica: Polycythemia Vera : the natural history of 1213 patients followed for 20 years. Ann Intern Med 1995, 125:656–664
2. Pearce JMS, Chandrasekera CP, Ladusans EJ. Lacunar infarcts in polycythemia with raised packed cell volume . BMJ 1983; 287:935-36
3. Meng R, Zhou J,Ji XM et al. The diagnosis and treatment of polycythemia rubravera manifesting as acute cerebral stroke. ZhonghuaNeiKeZaZhi 2006; 45(5): 366-8.
4. Silver RT. Treatment of Polycythemia Vera with recombinant interferon alpha (rIFNalpha) or imatinibmesylate. Curr Hematol Rep 2005; 4:235.
5. Finazzi G. Risk stratification , staging and treatment of patients with Polycythemia Vera : Italian and EuropianCollaberation on low dose aspirin in Polycythemia Vera .SeminThrombHemost. 2006;32:276-82.
6. Zoraster RM, Rison RA. Acute embolic cerebral ischemia as an initial presentation of Polycythemia Vera : a case report. J Med Case Rep. 2013 May 19;7:131.
7. Siddiqu M R, Mondol B A. Ischaemic Stroke as A Presenting Feature of Polycythemia RubraVera - A Case Report .AKMMC J 2013: 4(1): 45-47