## CASE REPORT

# ACUTE EMBOLIC CEREBRAL ISCHEMIA AS AN INITIAL PRESENTATION OF POLYCYTHEMIA VERA

Chirag M Chhatwani<sup>1</sup>, Ashok K Gagiya<sup>2</sup>, Ashish R Patel<sup>1</sup>, Chetan D Maniya<sup>1</sup>

Author's Affiliations: <sup>1</sup>Postgraduate Resident, <sup>2</sup>Associate Professor, Department of Medicine, SMIMER, Surat, Gujarat Correspondence: Dr Chirag M Chhatwani Email: chirag7166@gmail.com

# ABSTRACT

Patients with Polycythemia vera (PV) are at high risk for vaso-occlusive events including cerebral ischemia. Ischemic stroke may be the first presenting symptom of PV in 15% or more of those affected. It had been previously assumed that cerebral ischemic events were due to increased blood viscosity and platelet activation within the central nervous system arterial vessels. However, there are now a few isolated case reports of probable micro-embolic events originating from outside of the brain.

Case report: A 45-year old man presented with left sided hemiperesis (recovered within 12 hours) in our Medicine OPD. Hematologic investigation revealed a hyperviscous state (Hemoglobin 21.9gm% and PCV 66%). Acute infarction in right corona radiata and basal ganglia was found in magnetic resonance imaging(MRI) of brain. Although unusual, acute embolic cerebral ischemia may be an initial presentation of PV. The etiology of stroke in polycythemic patients is likely to be multifactorial. All clinicians involved in the care of stroke patients should be aware of the association of PV and ischemic stroke.

Keywords: Polycythemia vera, Cerebrovascular stroke, Embolic

# INTRODUCTION

Polycythemia vera is the clonal stem cell proliferation of red blood cells, white blood cells and platelets. The clinical manifestations of polycythemia vera are mainly due to increased productions of red blood cells and platelets. Increased production of white blood cells does no lead to any significant effects.1 Clinical presentation of polycythemis vera can be varied. Some people are diagnosed incidentally on hematological work while others can present with complications like stroke, deep vein thrombosis and pulmonary embolism.1 Ischemic stroke may be the first clinical presentation in 15% of those affected with the disease.2 It has been assumed that the reason for ischemic stroke in majority of cases of polycythemia vera is platelet activation and thrombus formation in cerebral arteries.3,4 Some studies have also suggested embolism as the cause of cerebral stroke in cases of polycythemia vera. Here we are presenting a case report polycythemia vera who presented as transient ischemic attack.

#### CASE REPORT

A 45-year old male presented to the Emergency Department with history of acute onset of left sided weakness involving both upper and lower limb. There was no history of sensory dysfunction. Patient

also had intermittent complain of headache and diminission of vision. Patient's brother also had similar complains of headache and diminission of vision. There was no history of diabetes or hypertension.

On examination, vitals were stable and blood pressure was normal. Cranial nerves 2–12 were all intact, speech was fluent without aphasia or dysarthria. Motor examination shows decrease power and tone in left side. Deep tendon reflexes were exaggerated over left side. There was no sensory dysfunction. Respiratory and cardiovascular system examination was unremarkable.

Laboratory investigations revealed Hemoglobin of 21.9, PCV was 66, TLC was 6300, Platelet count was 1.72, RBC count 7.03, MCV 93, MCH 31, RDW 17, Reticulocyte Count was 0.5%, LDH 600 and Uric acid was 4.4. Chest X ray and ECG were normal. MRI Brain revealed small acute infarct in right corona radiate and basal ganglia.

Patient improved completely within 12 hours and thus was diagnosed of having Transient ischemic attack. Detailed investigations were carried out to find out the etiology.

Serum erythropoeitin level was 3.64(3.7-30).CT Abdomen and 2D ECHO was normal.PFT was normal. Karyotype was normal and DNA PCR study for JAK2 mutation was negative. Bone marrow examina-

tion was suggestive of hypercellular marrow with increase in tri-lineage haematopoiesis. Thus a diagnosis of Polycythemia vera presenting as acute embolic cerebral ischemia was made.

Patient was given IV fluids and Phlebotomy done. Gradually patient's Hemoglobin and PCV came to 14.5mg% and 45% respectively.

### **DISCUSSION & CONCLUSION**

Incidence of Polycythemia vera is around 0.6- 1.6 per million population. It is most common in the age group of 50-70 years.<sup>5</sup> Increase in hematocrit the major factor responsible of increase in viscosity leading to decreased cerebral blood flow. <sup>4,6</sup> Local effect of increased hematocrit, increased viscosity, stagnation of blood and thrombocytosis lead to thrombus formation which is the most common mechanism of cerebral ischemia in polycythemia vera. <sup>4,6</sup> Another proposed mechanism is the emboli formation of which there are only few published case reports. <sup>7,8</sup> In our case patient presented with acute embolic cerebral ischemia and responded well with hemodilution and phlebotomy.

## REFERENCES

- Polycythemia vera facts. FS13. Leukemia & Lymphoma Society Web site. June 2012 [cited 2016 April 3].
- Zoraster RM, Rison RA. Acute embolic cerebral ischemia as an initial presentation of polycythemia vera: a case report. Journal of Medical Case Reports 2013; 7:131. doi:10.1186/1752-1947-7-131.
- Landolfi R, Marchioli R, Kutti J, Gisslinger H, Tognoni G, Patrono C, Barbui T. European Collaboration on Low-Dose Aspirin in Polycythemia Vera Investigators: Efficacy and safety of low-dose aspirin in polycythemia vera. N Engl J Med 2004; 350 Suppl 2:114–124.
- Spivak JL. The optimal management of polycythemia vera. Br J Haematol 2002; 116 Suppl 2:243–254.
- Besa EC, Woermann UJ: Polycythemia vera. Emedicine 2012 [cited 2016 April 04]. Available from: http://emedicine.medscape.com/article/205114-overview
- Spivak JL. Polycythemia vera: myths, mechanisms, and management. Blood 2002; 100 Suppl 13:4272–4290.
- Zimmerman C, Walther EU, von Scheit W, Hamann GF. Ischemic stroke in a 29-year-old man with left atrial spontaneous echoes and polycythemia vera. J Neurol 1999; 246 Suppl 12:1201–1203.
- Segura T, Serena J, Teruel J, Dávalos A. Cerebral embolism in a patient with polycythemia rubra vera. Eur J Neurol 2000; 7 Suppl 1:87–90.