

## CASE REPORTS

# Splenic Infarction in Polycythaemia Vera : Can the Spleen be Saved?

Sonali Inamdar\*, Shashikala Sangle\*\*, Nitin N Bote\*, Nitin Nagargoje\*

### Abstract

Polycythaemia vera is associated with thrombotic phenomenon due to hyperviscosity of blood. Splenic infarction with splenomegaly is a catastrophic complication usually requiring splenectomy. We describe a case of splenic infarction as an initial manifestation of polycythaemia vera which was treated with serial phlebotomies and hydroxyurea alone, without the need for surgery.

### Introduction

Polycythaemia vera is a clonal disorder involving a multipotent haematopoietic progenitor cell in which phenotypically normal red cells, granulocytes, and platelets accumulate in the absence of a recognisable physiologic stimulus. There is an increase in the number of circulating RBCs per volume of blood reflected as an elevated haemoglobin and haematocrit levels.

Thrombotic events are common due to hyperviscosity of blood.<sup>1</sup> Stroke, transient ischaemic attacks, deep venous thrombosis, myocardial infarction, retinal artery or vein occlusion and Budd-Chiari syndrome are the commonly seen complications of thrombosis. Splenomegaly is usually seen as a consequence of extramedullary haematopoiesis.<sup>1,2</sup> Splenic infarction with splenomegaly is an infrequent thrombotic manifestation seen in polycythaemia vera.<sup>3</sup> Painful splenomegaly with infarction or compression of surrounding viscera are the presently accepted indications for a splenectomy.<sup>4</sup>

### Case Report

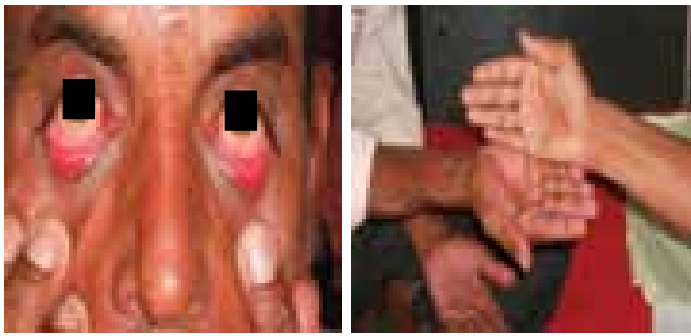
A 45 yr old male farmer was admitted for pain in the left hypochondriac region since 2 months. It was dragging in nature, increasing in intensity on inspiration. There was no history of blunt trauma, fever, haematuria. He was a known hypertensive since 10 yrs on regular antihypertensives. On examination, he was afebrile, pulse rate 84/min, blood pressure 110/70 mmHg and respiratory rate 22/min.

There was no icterus/lymphadenopathy. However, he had prominent facial flushing along with congested conjunctiva. Also, there was diffuse erythema of his palms (Figure 1).

On systemic examination, he had splenomegaly of 16 cm below the left subcostal margin. It was firm, non tender with no splenic rub. Other systemic examination was normal.

His investigations revealed haemoglobin of 20 gm% and a haematocrit of 64% along with an increased red cell count ( $9.21 \times 10^4$  /ul). There was leucocytosis and thrombocytosis too. Peripheral blood smear was hypercellular on low power microscopy and as a result, it was not possible to delineate the type of cells. Chest X-ray, ECG and urine examination were normal.

\*Resident Doctor, \*\*Associate Professor, Department of Medicine, BJ Medical College and Sassoon General Hospitals, Station Road, Pune-411001, Maharashtra.  
Received: 14.02.2013;  
Accepted: 02.09.2013



**Fig. 1 : Conjunctival congestion, facial flushing and palmar erythema**

The patient's USG abdomen showed a splenomegaly of 18 cm with a 8 cm x 4.4 cm sized heterogeneous hypoechoic lesion at the mid pole of spleen, likely a splenic infarct. Liver and kidneys were normal.

CT abdomen revealed moderate to severe splenomegaly. Craniocaudal span measured 20cm. A wedge shaped, subcapsular hypodense nonenhancing area with irregular but well defined margins involving the posterosuperior parenchyma was present suggestive of an infarct (Figure 2).

Bone marrow aspiration showed hypercellularity with trilineage growth. Serum erythropoietin was normal - 6mIU/ml (normal range 3.7– 29.5mIU/ml). JAK 2 gene v617f mutation was positive.

A final diagnosis of massive splenomegaly with a large splenic infarct in a case of primary polycythaemia (Polycythaemia vera) was done. The patient was treated with weekly phlebotomies and tablet Hydroxyurea 500 mg BD. His serial haemogram showed a response to treatment with the haemoglobin dropping to 14 gm% at the end of 4 weeks (Table 1). A repeat USG abdomen showed regression of the splenic size along with subsequent shrinking of the infarct size. The patient's facial plethora also decreased markedly.

Thus, our patient of large splenic infarction with splenomegaly due to Polycythaemia vera was treated successfully with phlebotomy and a myelosuppressive agent (Hydroxyurea) alone, without the need of splenectomy.

## Discussion

Thrombotic complications occur commonly in Polycythaemia vera. In 12% to 49% they occur as a presenting symptom. In 14% of cases, thrombotic events precede diagnosis. The central nervous system is the most frequent site of thrombosis leading to strokes or TIAs. Arterial thromboses are more commonly seen than venous. Venous thromboses are usually cerebral or intraabdominal, found more in women. Haemorrhagic complications can also occur due to vascular stasis and thrombocytosis, affecting mainly the CNS and GI systems. Splenic infarction

due to thrombosis is infrequent.<sup>1,2</sup>

Thromboembolic infarction of the spleen can cause nonspecific signs and symptoms, including abdominal pain, fever, and tachycardia.<sup>5</sup> The typical appearance of a splenic infarction is peripheral, low-attenuation, wedge-shaped infarcts. In some cases, splenic infarcts may appear on CT as large hypodense lesions.<sup>3</sup>

Phlebotomy is the best initial treatment for Polycythaemia vera as it ensures a rapid way to reduce the increased red blood cells down to normal levels. Removal of 500 cc blood 1-2 times a week to target a haematocrit of 45% in males and 42% in females is recommended.

It is however, associated with an increased risk of thrombosis; especially for patients older than 70 years, those with a history of thrombosis, and those requiring an increased frequency of phlebotomy. Concurrent treatment with myelosuppressive agents decreases the risk of thrombotic complications.<sup>6</sup>

Hydroxyurea is usually well tolerated and cheap and has been proven effective in many studies for the prevention of thrombohaemorrhagic complications. It is usually used in conjunction with phlebotomy as was done in our case. There is however, a perceived risk of progression to acute leukaemia. Several recent, large studies have given this drug a new lease of life in which the risk of leukaemia is placed in perspective to demonstrate that hydroxyurea still remains the drug of choice in patients with polycythemia vera.<sup>7</sup>

Other treatment options for Polycythaemia vera include interferon alpha and anagrelide (cyclic adenosine monophosphate phosphodiesterase inhibitor).

Polycythaemia vera is a condition that surgeons do not commonly encounter. Advances in medical management have largely led to avoidance of surgical intervention in most patients. Indications and timing of splenectomy have been the subject of debate since the disease was first described in the late 19th century. Painful splenomegaly with infarction or compression of surrounding viscera are presently accepted indications for surgery.<sup>4</sup>

Park EJ et al described a case report of massive splenic infarction in Polycythaemia vera, treated with phlebotomy and a myelosuppressive agent alone; as seen in our case.<sup>6</sup> Another case of massive splenic infarction with subsequent liquefaction is described by D. Beckett et al, which was managed medically as well.<sup>8</sup>

## Follow up

Our patient has been thus successfully treated with phlebotomy and hydroxyurea and has maintained his



**Fig. 2 : CT abdomen – Moderate to severe splenomegaly with a wedge shaped subcapsular splenic infarct.**

haemoglobin level at around 13-14 gm% No further complications of Polycythemia have arisen and he is on regular follow up.

## Conclusion

Splenic infarction due to polycythaemia vera generally entails a need for surgery. A trial of phlebotomy with a myelosuppressive agent can obviate the need of a splenectomy and its associated risk of infections and long term morbidity.

## Acknowledgements

We express our sincere thanks to haemato-oncologist Dr. Sameer Melinkeri and pathologist Dr. Aditi Dastane for their invaluable contributions towards the management of this case.

**Table 1: Serial hemogram (\*denotes phlebotomy done on each day)**

Serial hemogram	15/9*	20/9*	27/9*	3/10*	18/10*
Hb gm/dl	20	17.7	16.7	15.9	14.1
Hematocrit (%)	64.2	68.7	69.3	54.4	56.2
MCV (fL)	69.7	83.6	92.2	80.2	90.3
MCHC g/dL	31.2	25.7	24	29.2	26
MCH (pg)	21.7	21.5	22.2	23.4	23.5
MPV (fL)	10.8	8.3	8.3	7.7	8.2
PLATLET (/uL)	522000	377000	341000	244000	102000
RBC ( $\times 10^6$ ) / uL	9.21	8.21	7.51	6.78	6.21
RDW (%)	27.9	19.1	20.3	21.4	23.1
WBC ( $\times 10^3$ ) / uL	18.7	14	12	8.6	7.7

## References

1. Gruppo Italiano Studio Policitemia. Polycythemia vera: the natural history of 1213 patients followed over 20 years. *Ann Intern Med* 1995;123:656-64.
2. Gumina RJ, Foley DA, Tefferi A. Polycythemia vera—a case report and discussion on pathogenic mechanisms of increased thrombosis. *Angiology* 2002;53:587-91.
3. Balcan I, Seltzer SE, Davis S, Gellen S. CT patterns of splenic infarction: a clinical and experimental study. *Radiology* 1984;151:723-729.
4. Logan MS, Watson CM, Nottingham JM. Symptomatic splenomegaly in polycythemia vera: a review of the indications for splenectomy and perioperative considerations. *Am Surg.* 2009;75:363-8.
5. O'Keefe JH Jr, Holmes DR, Schaff HV, et al. Thromboembolic splenic infarction. *Mayo Clin Proc* 1986;61:967-972.
6. Park EJ, Han JY, Kim JY, Joo DH, Kim HS, Lee KS. A Case of Multiple Splenic Infarction in Polycythemia Vera treated with Phlebotomy alone. *Korean J Med* 1998;55(6):1107-1107.
7. Dingli D, Tefferi A. Hydroxyurea: The drug of choice for polycythemia vera and essential thrombocythemia. *Curr Hematol Malig Rep.* 2006;1:69-74.
8. Beckett D, Miller C, Ferrando JR, Banerjee AK. Polycythaemia vera presenting as massive splenic infarction and liquefaction. *The British Journal of Radiology*, 2004;77:876-877.