

Case Report

Subacute Intestinal obstruction associated with a case of Polycythemia Vera — a rare combination

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Abstract

Primary Polycythemia Vera (PV) is difficult to diagnose due to its varied and atypical presentation. It is a myeloproliferative disorder which is managed conservatively.

We submit a case report of a patient who was admitted in surgery with subacute intestinal obstruction due to compression by a massive splenic cyst. The investigations showed PV causing splenomegaly.

Introduction

Primary Polycythemia Vera (PV) is a myeloproliferative disorder wherein the red cell mass increases, leading to a rise in haematocrit and hyperviscosity of blood which may result in vascular thrombosis.¹ There may be an associated increase in white blood cell and platelet counts too. Secondary causes of polycythemia are smoking, chronic pulmonary disease and renal disease and they should be ruled out before making a definitive diagnosis of PV.²

Presenting features are high haemoglobin and haematocrit along with splenomegaly, pruritus, thrombotic events (e.g. transient ischaemia and infarcts), gastrointestinal disturbances, headache, malaise, breathlessness or plethora.³

Subacute intestinal obstruction is not associated with this disease and in this case it was secondary to extra mural compression of intestines by the massive splenic cyst.

Case Report

A 45 years, non-smoker female came to surgical emergency with longstanding left hemiparesis, intermittent nausea, vomiting and epigastric pain of one month duration. She also had heaviness in left hypochondrium and pruritus for 3 months. She had an old contrast enhanced computerized tomography (CECT) scan of cranium documenting right-sided parieto-occipital non-haemorrhagic infarct (Figure-1).

On examination her abdomen was nontender but distended. There was a 10 x 10 cm lump in the left hypochondrium which was firm and extended to the epigastrium and the left lumbar region. It was dull on percussion and there was no bruit on auscultation and abdominal sounds were sluggish.

The patient was managed conservatively by

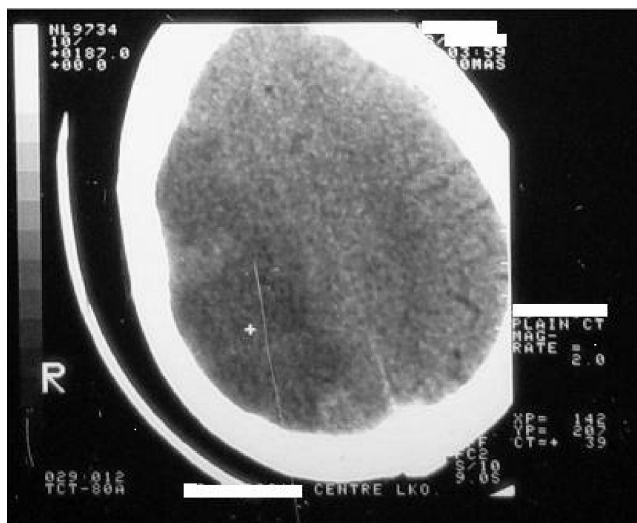


Figure-1: Old CT scan cranium demonstrating a non-haemorrhagic infarct.

intravenous fluids and antibiotics along with gravity assisted nasogastric tube aspiration. Haematological investigations showed a haemoglobin of 19.4 g/L, haematocrit of 0.6, Total Red Blood Count was $5.90 \times 10^{12}/L$, total leukocyte count was $14.8 \times 10^9/L$ with differential leukocyte count of, neutrophils 63%, lymphocyte 35%, monocyte 2%, eosinophil 0%, and basophil 0%, platelet count was $680 \times 10^9/L$.

Chest X-ray was normal. Plain x-ray abdomen showed multiple fluid levels. Ultrasound scan of abdomen demonstrated a thick walled cystic lesion in the splenic area with normal liver and kidneys. CECT scan of abdomen showed a large splenic cyst approximately 15cm x 10cm x 10 cm in size, displacing and compressing the surrounding structures, especially the upper gastrointestinal tract (GIT). Splenic and portal veins were not dilated on CECT scan which eliminated portal hypertension as a probable cause of splenomegaly (Figure-2).

An ultrasound guided pigtail catheter placed in the cyst yielded 1500 mL of fluid which on microscopic examination showed pus cells along with degenerated RBCs. No malignant cells were found and bacteriology was negative. Pig tail catheterization was therapeutic since her obstructive symptoms gradually resolved over the next three days which further strengthens the assumption that the obstruction was

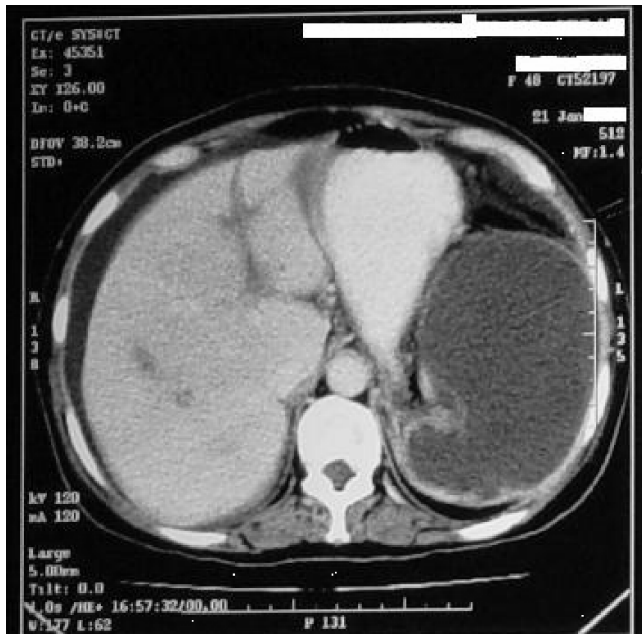


Figure-2: CECT scan of the abdomen showing a splenic cyst compressing and displacing the surrounding structures.

caused due to the cyst and resolved following cyst decompression.

Persistently maintained oxygen saturation (SpO₂) between 85- 90% range ruled out secondary polycythemia indirectly. Serum Erythropoietin level measurement could not be done due to economical reasons. Bone marrow aspiration was done once the general condition of the patient improved and showed marked erythroid hyperplasia with increase in

myeloid series cells.

Splenic abscess was excluded after clinical correlation of symptoms and findings of the CECT scan. Splenic infarct due to splenic vein thrombosis was the most probable cause of cyst formation in this patient. The patient was managed by hydroxyurea therapy and phlebotomy along with aspirin and antihistamines and other supportive drugs but she was non-compliant and succumbed after 8 months of initial diagnosis.

Comment:

Two major and two minor criteria needed to support the diagnosis of PV were present in this patient (as recommended by the World Health Organization committee).² Chromosomal analysis might have helped in clinching the diagnosis but could not be done due to financial constraints.³ CECT cranium documenting previous thrombotic events with a high haemoglobin level raised our suspicion towards PV.

Though the PV itself was not a direct cause of intestinal obstruction but it is an indirect cause since the cyst formed due to PV led to the intestinal obstruction. This association is a rare finding and has not been reported earlier to the best of our knowledge.

References

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