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An International Multidisciplinary e-Journal
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Email: researchinspiration.com@gmail.com, publish1257@gmail.com
ISSN: 2455-443X
Vol. 1, Issue-III
June 2016
Impact Factor : 4.012

# MYELOPROLIFERATIVE DISORDER PRESENTING AS PORTAL AND MESENTERIC VENOUS THROMBOSIS - A CASE REPORT

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## ABSTRACT

*Myeloproliferative disorders (MPD) can present as unprovoked venous thrombosis, in particular abdominal venous thrombosis. The events of thrombosis can occur even in the presence of a normal peripheral cell counts. We report the case of a 32 year old male presenting with abdominal pain, sitophobia and non-tender splenomegaly. Investigations done revealed high platelet counts, gastroscopy showed large oesophageal varices, portal doppler showed normal liver echoes and cavernous transformation of portal vein, CECT abdomen showed chronic portal and superior mesenteric venous thrombosis. Bone marrow biopsy revealed increased megakaryocytic precursors and JAK2V617F mutation was positive. Hence a diagnosis of myeloproliferative disorder (essential thrombocythemia ) presenting as portal and mesenteric venous thrombosis was made. Patient underwent endoscopic variceal ligation followed by anticoagulation and Hydroxyurea therapy. He showed good response to treatment till date. This case highlights the importance of thorough*

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*screening for myeloproliferative disorders in patients presenting with portal and mesenteric vein thrombosis.*

**KEY WORDS**- Gastroscopy, Varices, Thrombocythemia

## INTRODUCTION

Thrombosis of Mesenteric and Portal Venous system are commonly encountered in day to day Gastroenterology practice. Myeloproliferative disorders (MPD) have been reported to be common prothrombotic cause of non cirrhotic, non malignant Portal Vein Thrombosis (PVT). Combined PVT and Mesenteric Vein Thrombosis (MVT) may present with a variety of acute symptoms and potentially fatal signs. A detailed prothrombotic work up is needed in such cases and management has to be tailored on case to case basis. We report such case of portal and mesenteric venous thromboses due to Essential thrombocythemia (ET), a Myeloproliferative disorder, from our Institute.

## CASE REPORT

A 32 years old male, presented with history of abdominal pain, fear of eating and weight loss for three months, no past history of GI bleeds, jaundice, no family history of liver disease or a hypercoagulable state. On examination he had firm non tender splenomegaly 10 cms below left costal margin. His cardiovascular, respiratory and central nervous system were clinically normal. Investigations: complete blood count –leucocyte count of 7900 cells/cumm, hemoglobin -12.9gms/dl, platelet count -10,17,000 cells /cumm. Peripheral smear study showed normocytic, normochromic RBCs, much increased platelet counts. Blood sugar-112 mgs/dl, Blood urea-39 mg/dl, S. creatinine-0.9 mg/dl ,Total Bilirubin -1.1mg/dl, Direct -0.3mg/dl, SGOT -30, SGPT-28, T. protein-7.1,

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Albumin -3.7, PT (14.5/12) and aPTT (38.5/32). HBsAg- negative, Anti HCV- negative, HIV-negative, ECG- Normal, Chest x ray-normal study, ECHO cardiogram- normal. Gastroscopy: Esophageal varices gr 3× 3 columns. Portal Doppler- portal venous cavernoma, superior mesenteric vein thrombosis and Splenomegaly. CT angiogram done confirmed cavernous transformation of portal vein, SMV thrombosis, Inferior mesenteric vein partial thrombosis.(fig.1&2)

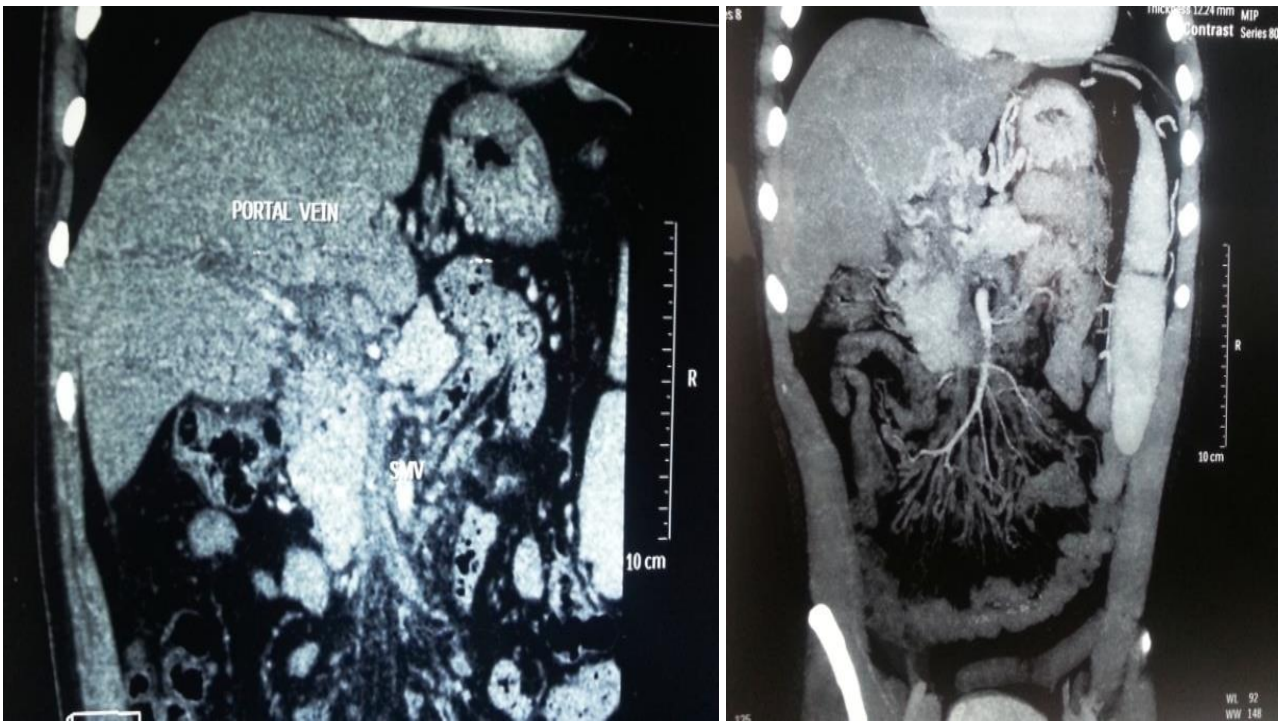


Fig 1&2- CT angiogram showing Portal cavernoma, superior mesenteric vein thrombosis& portosystemic collaterals

Bone marrow examination showed – marked increase in number of megakaryocytes with hyperlobated nuclei and eosinophilic cytoplasm, suggesting – Essential Thrombocythemia (fig.3) Hence JAK2 V617F mutation study was done. It was positive (fig.4).

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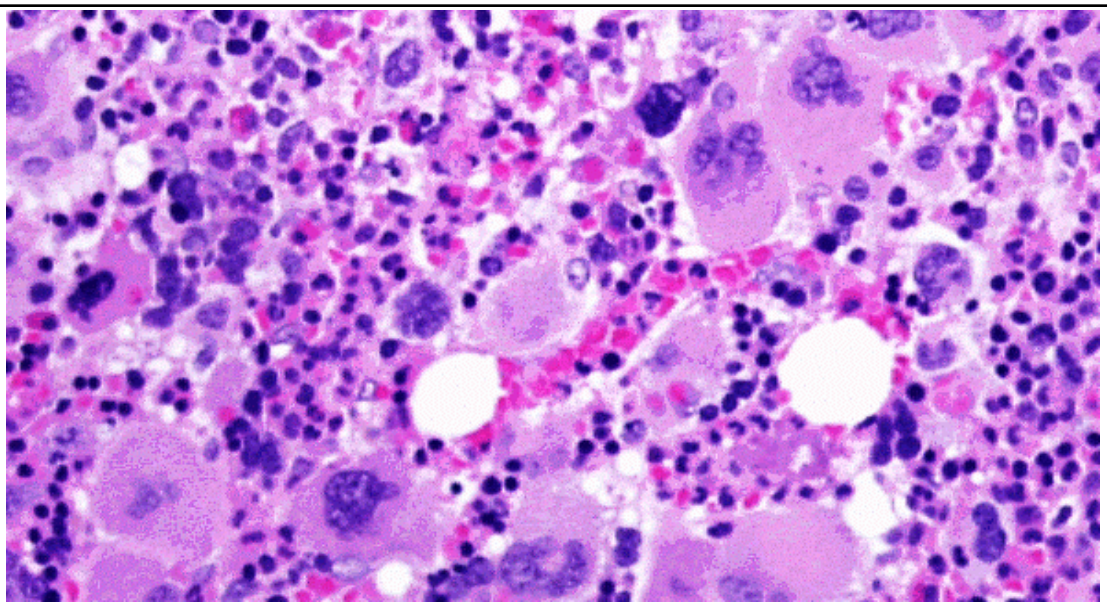

Fig 3- bone marrow biopsy showing increased megakaryocytes with hyperlobated nuclei &eosinophilic cytoplasm

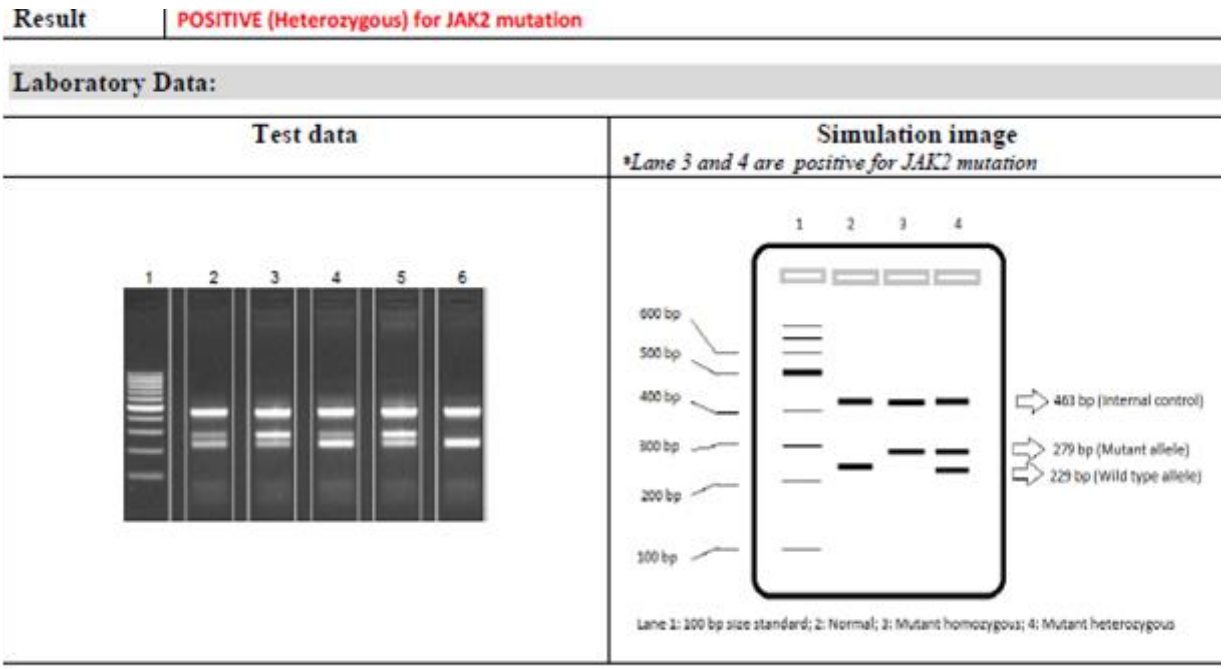


Fig 4- shows JAK2 V617F mutation analysis by PCR

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Hence a final diagnosis of: Myeloproliferative disorder-Essential thrombocythemia, presenting as chronic portal and mesenteric venous thromboses was made. Patient underwent endoscopic variceal ligation following which low molecular weight heparin, vitamin K antagonists, C. Hydroxyurea were started. Following that patients platelet count gradually normalized after 3 months and he did not have any adverse event till date.

## DISCUSSION

Essential thrombocythemia is a chronic myeloproliferative disorder affecting megakaryocytic cell lineage. Although half of the patients may be aymptomatic at presentation, remaining may present with vasomotor symptoms like headache, atypical chest pain, syncope, livido reticularis, transient visual disturbances etc. They can also manifest with thrombo-embolic complications like stroke, transient ischemic attacks, pulmonary embolism, retinal vessel occlusions, coronary artery ischemia, Hepatic or portal vein thrombosis, deep vein thrombosis and digital ischemia.

Around 50% of patients with ET have shown JAK2V617F mutation. But the presence of this mutation currently does not differentiate ET, polycythemia vera and Chronic MyeloFibrosis.[1,2] .Yet, ET can be diagnosed by combined use of Bone marrow histopathology and JAK2V617F mutation detection. Our patient had the typical clinical picture, with positive bone marrow biopsy suggesting ET supported by JAK2V617F mutation.

ET is associated with abdominal vein thrombosis, the incidence being 11-25% [3,4]. Large vessel thrombosis as complication of ET has been reported in 18-51% of cases in various series. [5,6].The prevalence of PVT was 1% in general population according to a study conducted at Sweden[7].The causes of PVT include Inhertied and Acquired Prothrombotic states like Primary

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myeloproliferative disorders, Intra-abdominal inflammation, Portal vein injury , liver cirrhosis, Budd-Chiari syndrome, Malignancies, Pregnancy and oral contraceptives. The incidence of MPD is reported to be 37% in patients with non-cirrhotic, non malignant PVT [8].

Acute PVT can present with nausea, vomiting, abdominal pain, acute onset ascites, hematochezia and splenomegaly. Chronic PVT is usually asymptomatic but may present with symptoms of portal hypertension like GI bleed and hypersplenism.

Laboratory investigations show normal liver function except if PVT occurs in a patient with cirrhosis and a rise in alkaline phosphatase in portal biliopthy. In ultrasound imaging, the liver is normal with solid echoes within portal vein in PVT, but may show nodules in liver and portal cavernoma in chronic PVT. CECT abdomen may show the extension of thrombus, evidence of bowel infarction and status of adjacent organs and contrast CT can differentiate a bland and a malignant thrombus. Magnetic resonance venogram is valuable in determining the resectability of neoplasm involving the portal venous system and follow-up after therapeutic shunt procedures.

Treatment of PVT depends on the precipitating event like prothombotic state, age and extent of the thrombus and status of the liver. In cases with normal liver function, acute and symptomatic PVT can be treated initially with Low Molecular Weight Heparin followed by oral anticoagulants for three to six months to prevent total obstruction of vein by thrombus and cavernous transformation. Chronic oral anticoagulation for lifelong has been recommended for patients with hypercoagulable disorders or if thrombus had extended to mesenteric vein. In patients with cirrhosis, anticoagulants are not given routinely except in pre transplant cases, associated hypercoagulable state or mesenteric vein extension. In cases of MPD with upper GI bleed from eso gastric varices, endoscopic band

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ligation /sclerotherapy are considered mandatory before starting anticoagulant therapy. In refractory variceal bleeding, Trans jugular Intra hepatic Portosystemic shunt (TIPS) placement is recommended. Chronic anticoagulant therapy has not found to cause a significant increase in risk and severity of variceal bleeding as reports in few studies.[9] In PVT associated with MPDs, hydroxyurea or platelet specific lowering agents like Anagrelide can be given to reduce thrombotic complications further. Failure of anticoagulation therapy can be managed by thrombolysis into SMV or Portal vein or mechanical thrombectomy should be done.

Although MPDs are common than other acquired causes of PVT (excluding cirrhosis and cancer), only a few confirmed cases have been reported in Indian population. Though anticoagulants were considered risky in our patient, we started anticoagulants after EVL to prevent further thrombotic events which has proved beneficial and safe till now.

## CONCLUSION

Portal vein thrombosis is rather easily diagnosed in modern day diagonostic armamentarium but yet very difficult to treat. A thorough search for inherited and acquired thrombotic states is needed in all patients with PVT before deciding the treatment protocol to avoid risk of recurrent thrombotic events.

## BIBLIOGRAPHY

1) Michiels JJ, Berneman Z, Schroyens W, HebedaK, LamK, De Raeve H.The 2001 WHO and updated European clinical and pathological criteria for the diagnosis, classification and staging of Philadelphia chromosome-negative chronic myeloproliferative disorders. Semin Thrombin Hemost2006 Jun;32(4 Pt 2):307-40
2) Tefferi A, Pardanani A. Mutation screening for JAK2V617F: when to order the test and how to interpret the results. Leukemia 2006; Res 108:3472–3476
3) Elliott MA, Tefferi A. Thrombosis and haemorrhage in polycythaemia vera and essential thrombocythaemia. Br J Haematol 2005;128:275–290
4) Johnson M, Gernsheimer T, Johansen K. Essential thrombocytosis: underemphasized cause of large-vessel thrombosis. J Vasc Surg 1995; 22:443–449.

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5) Kessler CM, Cline HG, Havlik RJ. Uncontrolled thrombocytosis in chronic myeloproliferative disorders. Br J Hematol 1982;50:157-60
6) Randi ML, Stocco F, Rossi C, Tison T, Girolami A. Thrombosis and hemorrhage in thrombocytosis: evaluation of a large cohort of patients (357 cases). J Med 1991; 22:213-223.
7) Ogren M, Bergqvist D, Bjorck M, Acosta S, Eriksson H, Sternby NH, Portal vein thrombosis: Prevalence, patient characteristics and lifetime risk: A population study based on 23 796 consecutive autopsies. World J Gastroenterol. Apr 7, 2006; 12(13): 2115-2119
8) Kiladjian JJ, Cervantes F, Leebeek FWG, Marzac C, Chevret S, Cassinat B,et al. Role of JAK 2 mutation detection in Budd-Chiari syndrome (BCS) and portal vein thrombosis (PVT) associated to MPD. Blood,2006;108:116a
9) Malkowski P, Pawlak J, Michalowicz B, Szczerban J,Wroblewski T,Leowska E,et al, Thrombolytic treatment of portal thrombosis.Hepatogastroenterology, 2003;50:2098

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