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PORTAL VEIN THROMBOSIS DUE TO ESSENTIAL THROMBOCYTOSIS

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Abstract : Portal vein thrombosis (PVT) is the blockage or narrowing of the portal vein by a thrombus. Portal vein thrombosis (PVT) is considered to be a frequent complication of liver cirrhosis. It is relatively rare and has been linked with the presence of an underlying liver disease or prothrombotic disorders. . It is not clear whether PVT is a consequence of severe liver disease or a factor which aggravate underlying liver disease or both. We report an interesting and rare case of PVT due to Essential Thrombocytosis management, difficulties we encountered and how we overcame them.

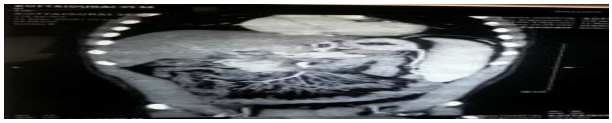
Keyword :PVT, Essential Thrombocytosis, prothrombotic

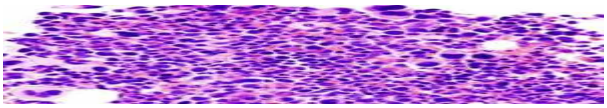
INTRODUCTION

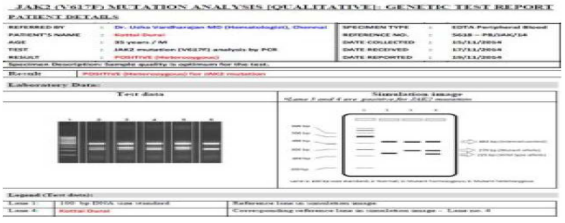
Portal vein thrombosis (PVT) is occlusion of portal vein or its branches by blood clot. This is encountered in a various clinical settings like myeloproliferative disorders, cirrhosis of liver, provoking factors like intra-abdominal septic focus, pancreatitis or cancer1. It is not clear whether PVT is a consequence of severe liver disease or a factor which aggravate underlying liver disease or both. Liver transplantation has altered the prognosis of patients with cirrhosis but presence of PVT can exclude a patient from transplant list due to low post transplant survival in them. There is no clear consensus regarding management of PVT in various clinical settings and initiation of anticoagulation/thrombolytics, monitoring them and duration of treatment still remain a grey zone. We report an interesting and rare case of PVT due Essential Thrombocytosis management, difficulties we encountered and how we overcame them.

CASE REPORT

A 32 years old male, presented with history of abdominal pain, fear of eating and loss of weight for 3 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 was clinically normal. Investigations revealed a 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 was 112 mgs/dl, Blood urea 39 mg/dl, Serum creatinine 0.9 mg/dl ,Total Bilirubin 1.1mg/dl, Direct 0.3mg/dl, SGOT 30, SGPT 28, Total protein 7.1, Albumin 3.7, INR 1.13 and aPTT (38.5/32). HBsAg negative, Anti HCV negative, HIV negative, ECG was normal, chest x ray was normal, ECHO cardiogram was also normal. OGD Scopy revealed esophageal varices grade 3× 3 columns. Portal Doppler showed cavernous transformation of portal vein, thrombosis of SMV and splenomegaly. CT angiogram done confirmed cavernous transformation of portal vein, superior mesenteric vein thrombosis, inferior mesenteric vein partial thrombosis. Bone marrow examination done showed marked increase in number of megakaryocytes with hyperlobated nuclei and eosinophilic cytoplasm, suggesting ESSENTIAL THROMBOCYTHEMIA, and JAK2 mutation (jannus kinase) was positive. Hence we came to a final diagnosis of myeloproliferative disorder Essential thrombocythemia, presenting as chronic portal and mesenteric venous thromboses. Patient underwent endoscopic variceal ligation following which low molecular weight heparin, vitamin K antagonists and hydroxyurea were started.







DISCUSSION:

Portal vein thrombosis refers to development of thrombus within extra-hepatic portal venous system draining into the liver. Anatomical classification **TYPE-1** : main portal vein, beyond confluence of SV/SMV(splenic vein/superior mesentric vein), **TYPE-2** : extends into SMV but with patent mesenteric vessels, **TYPE-3** : diffuse thrombosis of splanchnic vessels + large collaterals, **TYPE-4** : diffuse thrombosis of splanchnic vessels + finer collaterals, Types 1& 2 manifest as variceal bleeding, Types 3 & 4 present with bowel ischemia3. *Voorhees et al. (1974), Webb et al. (1979)* said the majority of patients with PVT did not have underlying etiologies4,5. However various etiologies for PVT has been proposed and mentioned in the table given below. Broadly they are classified as systemic and local thrombophilic disorders6. Systemic are again subclassified as inherited and acquired. Multiple factors contribute to the development of venous thrombosis. Infant with infection of the umbilical vein in the absence of prothrombotic disorders infrequently go on to develop PVT. Blunt trauma and surgical procedures, generally, do not precipitate PVT unless there is an associated prothrombotic state. Patient usually manifest as acute or chronic PVT. Acute PVT may be asymptomatic and if symptomatic they will have abdominal pain, fever, nausea. If the thrombosis extends to involve SMV they will have abdominal pain associated with hematochezia and intestinal ischemia features. Chronic PVT may be asymptomatic and if they develop symptoms they will have well tolerated UGI bleeds, splenomegaly, hypersplenism, growth retardation and obstructive jaundice.

Inherited Disorders - High risk of thrombosis - Anti thrombin III deficiency - Protein C deficiency - Protein S deficiency Low risk for thrombosis - Factor V Leiden mutation - Factor II (PTHR A) mutation	Infection/Inflammation - Neonatal Omphalitis - Appendicitis - Pancreatitis - Cholecystitis - Perforated peptic ulcer - Tuberculous lymphadenitis
Acquired Disorders - Malignancy - Myeloproliferative disorders - Oral contraceptive pills - Pregnancy and post partum - Anti Phospholipid syndrome - Paroxysmal nocturnal hemoglobinuria	Portal vein injury - Surgical shunts - Splenectomy - Abdominal surgery - Liver Transplantation - Blunt trauma
Mixed Disorders - Hyperhomocysteinemia	Cancer of the abdominal organs Cirrhosis

On evaluation patients with cirrhosis, abdominal cancers, infectious focus.one should check for multiple, concurrent risk factors for thrombosis in all patients without advanced cirrhosis/cancer7. Consider investigations for Protein C& S, anti-thrombin deficiency only when a first degree relative is positive. The liver function is normal except if PVT occurs in a patient with cirrhosis. Patients with portal hypertensive biliopathy may show a rise in alkaline phosphatase. Liver is grossly normal in PVT, but may show atrophy and regenerative nodular hyperplasia, related to apoptosis and compensatory arterial vasodilation in chronic PVT. USG Abdomen may show portal cavernoma suggestive of chronic PVT. Usually associated with splenomegaly and collaterals. It may show solid echoes within portal vein and absent flow on pulsed doppler.

The diagnostic sensitivity and specificity for Colour Doppler Ultrasound (CDUS) in detecting portal vein thrombosis vary from 66% to 100%. CT Scan may show extension of thrombus, evidence of bowel infarction and status of adjacent organs. On CECT Abdomen bland thrombus is seen as a low density, non enhancing defect within portal veins while a tumor thrombus enhances following contrast administration. MR Venography is valuable in determining the resectability of neoplasm involving the portal venous system and follow-up after therapeutic procedures including surgical spleno renal and meso caval shunts. AASLD recommendations for treatment of PVT varies according to the clinical presentation. Acute PVT give anticoagulants for more than 3months. Start with LMWH and oral anticoagulants overlap for first 5-7 days until we get a desired INR. Then withdraw LMWH and continue OAC in pts with acute On evaluation patients with cirrhosis, abdominal cancers, infectious focus.one should check for multiple, concurrent risk factors for thrombosis in all patients without advanced cirrhosis/cancer7. Consider investigations for Protein C& S, anti-thrombin deficiency only when a first degree relative is positive. The liver function is normal except if PVT occurs in a patient with cirrhosis. Patients with portal hypertensive biliopathy may show a rise in alkaline phosphatase. Liver is grossly normal in PVT, but may show

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PVT due to permanent thrombotic risk factors not corrected otherwise and thrombus extension into mesenteric veins. Initiate antibiotics in all acute PVT. Recanalization rates as high as upto 83% in these patients. Major bleeding was seen only in around 5% patients. Chronic PVT screen all patients with chronic PVT for varices. Treat large varices with EVL followed by beta blocker therapy. Long term oral anti coagulants in patients with chronic PVT without cirrhosis and with permanent risk factors for venous thrombosis not corrected otherwise. In our case we started with LMWH overlap with Warfarin and then after getting therapeutic INR we then switched over to warfarin monotherapy for life long. As it is a case of Essential thrombocytosis we started him on Hydroxyurea also. He responded well and he is under our follow up.

CONCLUSION:

We reported this case because portal vein thrombosis presenting as essential thrombocytosis is very rare and the thrombosis extending to superior mesenteric vein is extremely rare.

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