

## Portal cavernoma as a cause of hematemesis in a young unmarried female: A case report

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### ABSTRACT

Thrombotic incidents are usually uncommon in unusual sites like portal vein and further in the adolescent age group which presents a potential diagnostic dilemma. Chronic non-cirrhotic non-malignant portal vein thrombosis (PVT) can present at any age group, usually as acute upper gastrointestinal (UGI) bleed. Diagnosis depends on imaging modalities involving Doppler, computed tomography (CT), magnetic resonance imaging, and portography. We report the case of a 20-year-old unmarried girl presented with multiple episodes of hematemesis. Ultrasonography abdomen and CT abdomen revealed PVT with portal cavernous transformation of the portal vein. Her prothrombin time and activated partial thromboplastin time were normal. Her connective tissue workup was normal. A diagnosis of PVT secondary to umbilical vein sepsis during infancy was made. Adolescent patients presented with acute hematemesis should not be neglected for some local cause of UGI bleed but should be evaluated for other rare causes like PVT, for early diagnosis and management to reduce feared complications.

**Key words:** Endoscopic variceal ligation, Portal vein thrombosis, Upper gastrointestinal.

According to Organ *et al.*, the prevalence of portal vein thrombosis (PVT), as seen on autopsy series, is 1% [1]. It is estimated that PVT is responsible for 5–10% of the overall cases of portal hypertension in the Western world, while in the Indian subcontinent, it is 40% [2]. In the Indian subcontinent, variceal bleeding occurs due to PVT, their prevalence being 80% and 20–30% in children and adults, respectively [3], whereas, in Japan and the Western world, 10–20% and 2–5% of all variceal bleed are due to PVT, respectively.

Portal cavernoma is characterized by the disappearance of the normal portal vein and its replacement by a network of portoportal collaterals. This subsequent transformation of an acutely thrombosed portal vein into portal cavernoma is known as chronic PVT. The PVT occurs due to hypercoagulable state (such as antiphospholipid antibodies [ALPA], antithrombin deficiency, factor V Leiden mutation, methylenetetrahydrofolate reductase polymorphism, myeloproliferative neoplasm, nephrotic syndrome, oral contraceptives, paroxysmal nocturnal hemoglobinuria, prothrombin gene mutation, protein C, protein S deficiency, and sickle cell disease), portal pyemia secondary to intra-abdominal infections, inflammatory disease (Behcet's disease, inflammatory bowel disease, and pancreatitis), complications of therapeutic interventions (hepatobiliary surgery, transjugular intrahepatic portosystemic shunt [TIPS] procedure, umbilical vein catheterization, peritoneal dialysis, and radiofrequency ablation of hepatic tumor) splenectomy, and secondary to impaired portal

vein flow (such as Budd–Chiari syndrome, cirrhosis, pancreatic cancer, and sinusoidal obstruction syndrome).

### CASE REPORT

A 20-year-old unmarried girl belonging to poor socioeconomic status was presented to our medicine outpatient department with seven episodes of hematemesis with abdominal pain for 2 days and malena for 1 day, with no prior history of similar illness in the past. She has no history of jaundice, chest pain, palpitation, syncope, dyspnea, fever, chronic abdominal pain, and pulmonary tuberculosis. There was no significant medical and surgical illness in the past. Her menstrual history was unremarkable. As per her attendant, she was delivered at home by some local health worker, and her mother also revealed the culture and customs of the application of cow dung over the umbilical cord stump.

On admission, she was afebrile, her lying down blood pressure was 80/50 mmHg, pulse rate was 110/min, and pallor was present. Abdominal examination revealed splenomegaly 3 cm below the costal margin with no ascites, as confirmed by abdominal ultrasound.

Her full blood count revealed anemia with hemoglobin 6.4 g/dl, total leukocyte count was normal, platelets were 1,20,000, hematocrit was 28.4% with liver function tests (LFT), and renal function tests within the normal range. Her electrocardiogram and chest X-ray PA view were insignificant. Her prothrombin time and activated partial thromboplastin time were normal. Her

screening for HIV and hepatitis B and C virus was negative. The upper gastrointestinal (UGI) endoscopy revealed 3 column Grade 3 esophageal varices and gastric-esophageal varices type I.

The ultrasound abdomen and portal vein color Doppler show normal size/shape/echotexture of the liver with moderate splenomegaly (17 cm), dilated splenic vein (10 mm in diameter) with splenic hilar collaterals, and multiple torturous anechoic vascular channels at porta hepatis with the venous flow. On color Doppler, the main portal vein was not seen separately, suggestive of portal cavernoma. The contrast-enhanced computed tomography abdomen revealed enlarged caudate lobe of the liver with splenomegaly (20.6 cm) and cavernous transformation of the portal vein (CTPV) with dilated splenic vein and superior mesenteric vein. The antinuclear antibody screening by immunofluorescence showed an insignificant titer (1:80 titer). The diagnostic workup for APLA was negative (absent lupus anticoagulant and anticardiolipin antibodies). The patient was managed conservatively with an intravenous proton-pump inhibitor, intravenous octreotide, and blood transfusion and was advised for endoscopic variceal ligation (EVL).

## DISCUSSION

CTPV is a reparative process to bypass the already thrombosed native portal vein. It can arise in the intrahepatic branch of the portal vein or main portal vein itself secondary to occluding and non-occluding thrombus and help in the continued flow of blood even in the presence of clot [4]. Recanalization of the thrombosed vein can occur as early as 6–20 days after the formation of acute PVT [5]. This compensatory phenomenon is not able to handle mesenteric and splenic inflow leads to the formation of pre-sinusoidal portal hypertension and cirrhosis.

Clinical presentation depends on the timing and progression of thrombosis and collateral formation. Most patients have no symptoms or signs until they present with acute UGI bleeding secondary to portal hypertension [6]. Diagnosis depends on imaging studies involving Doppler, CT, magnetic resonance imaging, and portography. The LFTs are normal to near normal. The liver biopsy usually shows no abnormalities; however, sinusoidal dilatation [7], as well as regenerative hepatocellular change or minimal portal fibrosis [8], can be seen. The etiology of PVT is highly diverse, but in our patient, the cause of PVT may be umbilical vein infection due to the presence of significant risk factor that is home delivery and application of cow dung over the umbilical cord was present. Thrombocytopenia is a well-known complication of portal vein cavernoma. Therefore, regular determination of platelets is essential to prevent hemorrhagic complication [9].

The treatment option varies from anticoagulant therapy, surgical reconstruction, reduction of portal pressure by TIPS, and recurrent bleeding secondary to portal hypertension which can be prevented with non-selective beta-blocker and EVL [10]. Balancing the increased risk of bleeding and thrombosis in the management of PVT is the therapeutic challenge. Therefore, a decision regarding anticoagulation should be made on an

individual basis [11,12]. Anticoagulation should be initiated in all patients to prevent the extension of thrombus and recurrence of new thrombus [9]. Based on the available data, anticoagulation therapy is not associated with an increased risk of severity of GI bleeding as compared with an absence of anticoagulation therapy [6]. Therefore, a patient without esophageal varices and with a strong risk factor for thrombosis may benefit from anticoagulation therapy. In our patient, conservative management was done, and anticoagulation therapy was not given due to the presence of esophageal varices and thrombocytopenia.

Anti-coagulation further prevents the consumption of platelets and thus prevents consumption coagulopathy and mortality associated with it [9]. Pregnancy-associated with prior known PVT may complicate pregnancy because pregnancy itself is a hypercoagulable state and predispose to thrombosis [9]. Prognosis in portal cavernoma patients is related mostly to underlying conditions, not to complications of portal hypertension [12,13]. The involvement of a superior mesenteric vein is an independent predictor of a poor outcome [14,15]. Therefore, early diagnosis and treatment prevent feared complications such as intestinal infarction and perforation, which lead to increased mortality.

## CONCLUSION

This case report represents the unusual scenario of acute hematemesis in young unmarried females due to PVT secondary to umbilical vein infection during infancy as evident from home delivery and application of cow dung over umbilical cord. Therefore, young patients presented with acute hematemesis should not be neglected for some local cause of UGI bleed but should not be evaluated for the other rare cause like PVT. It should be considered with clinical practice for early intervention.

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