A rare pathological feature of portal vein thrombosis complicated by portal hypertension and haematemesis

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ABSTRACT

Portal cavernoma is a rare cause of upper gastrointestinal bleeding resulting from complications of portal hypertension. We report the case of a 30-year-old woman admitted with haematemesis and splenomegaly. Imaging revealed a portal cavernoma associated with oesophageal varices. The patient received a blood transfusion, fluid resuscitation, and on discharge was put on propranolol for secondary prevention. Endoscopic ligation could not be performed due to service limitations. Diagnosis of portal cavernoma relies on imaging. Its management is complex in low-resource settings such as Niger.

Keywords: haematemesis, cavernoma, portal hypertension, Zinder, Niger.

Introduction

Portal cavernoma, or cavernous transformation of the portal vein, is a rare vascular anomaly characterized by the development of a network of tortuous venous collaterals that bypass an obstructed portal vein.[1,2] It causes non-cirrhotic portal hypertension.[3] The exact incidence is unknown, but increasing use of Doppler ultrasound and CT imaging has improved recognition.[4] The condition may remain asymptomatic for years and often presents through complications such as upper gastrointestinal bleeding caused by rupture of oesophageal varices.[4,5] Clinically, the picture is dominated by splenomegaly, hypersplenism, and signs of portal hypertension.[5] The diagnosis rests on imaging—mainly Doppler ultrasound and CT scan—which demonstrate the absence of flow in the main portal vein and the presence of multiple serpiginous collateral vessels.[1] We report a case of portal cavernoma discovered following haematemesis in a young woman with no previous medical history and managed in a resource-limited environment.

Case Report

A 30-year-old woman with no history of liver disease, surgery, or thrombophilia presented to the Hepatology and Gastroenterology Department of Zinder National Hospital with haematemesis associated with dizziness and fever. She reported no use of non-steroidal anti-inflammatory drugs, no chronic abdominal pain, and no known peptic ulcer disease.

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At admission, she was pale, febrile (38.5°C), tachycardic (120 bpm), and hypotensive (80/60 mm Hg). Physical examination revealed stage III splenomegaly according to Hackett's classification and melaena on rectal examination. Laboratory investigations showed microcytic hypochromic anaemia (Hb = 5.9 g/dL) and thrombocytopenia (67,000/mm³). The thick blood film was positive for *Plasmodium falciparum*, while liver and renal function tests and coagulation studies were normal.

Initial management consisted of intravenous crystalloids with saline at 5ml/kg/hour, fresh blood transfusion at 756 ml to a transfusion target of 8g/dL, and 80 mg of omeprazole daily. Antimalarial therapy orally by artemether was initiated.

Upper gastrointestinal endoscopy was performed after stabilization, which revealed grade III oesophageal varices without red signs. The latter, e.g., cherry-red spots, would warn of imminent bleeding from weak areas. Abdominal ultrasound showed a serpiginous vascular formation in the hepatic hilum, suggestive of a portal cavernoma (Figure 1). Colour Doppler confirmed a chaotic flow within the portal venous network (Figures 2–3). A CT scan demonstrated a portal cavernoma without a focal hepatic lesion, associated with homogeneous splenomegaly (Figure 4). No biliary dilatation or mass was noted. A diagnosis of portal cavernoma complicated by variceal bleeding in a non-cirrhotic patient was made. Due to limited resources, endoscopic variceal ligation was unavailable. Anticoagulation was not indicated due to the chronic, organized nature of the thrombosis, and further bleeding might have been precipitated. Weekly follow-up over one month showed progress with cessation of bleeding and improved haemoglobin (10 g/dL). The patient received 40mg propranolol long-term, oral iron for three months, and six-monthly follow-up visits. She also received oral iron supplementation for three months.

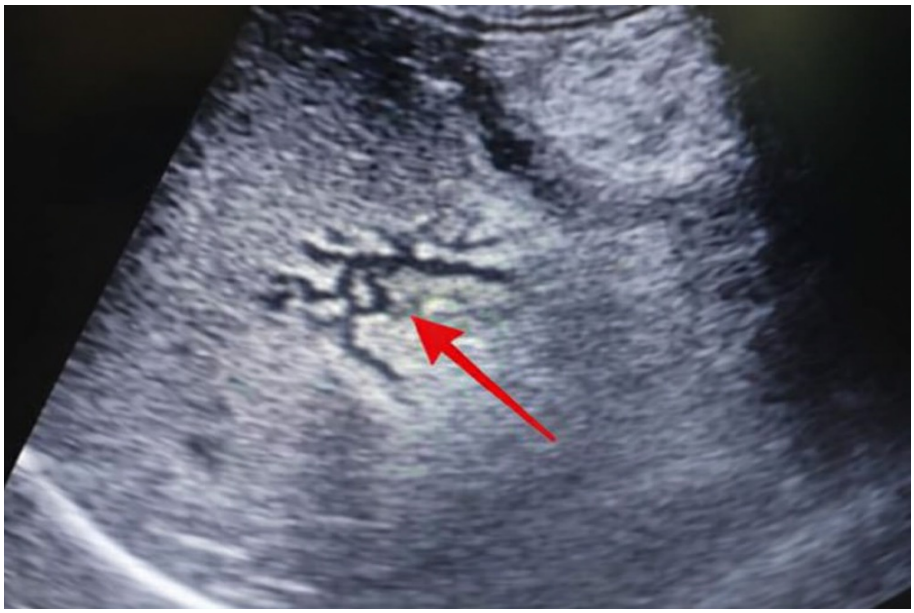

Figure 1. Abdominal ultrasound scan showing a serpiginous appearance at the hepatic hilum, in favour of a portal cavernoma.

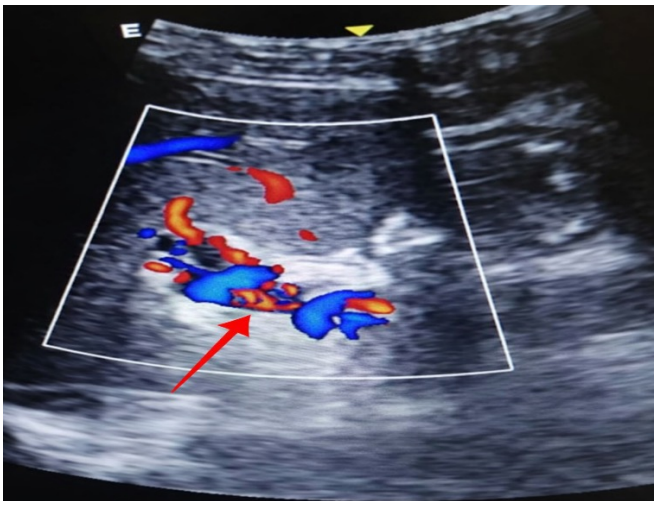

Figure 2. Doppler ultrasound illustrating a vascular malformation at the splenic vein level with heterogeneous flow, consistent with an ectopic vascular anastomosis.

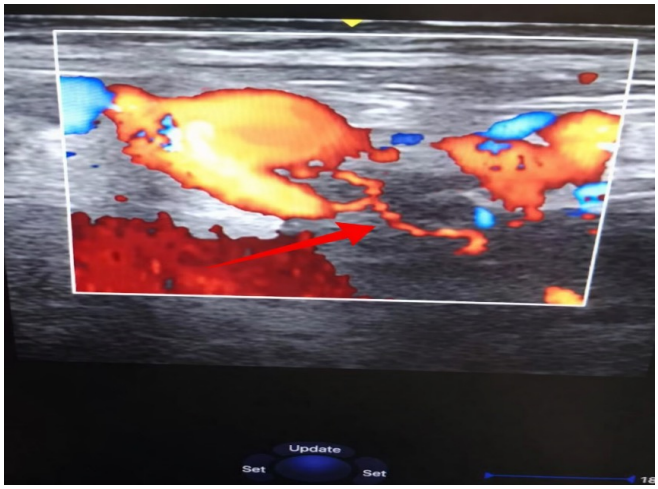

Figure 3. Doppler ultrasound showing the portal vein with a serpiginous appearance indicative of a portal cavernoma.

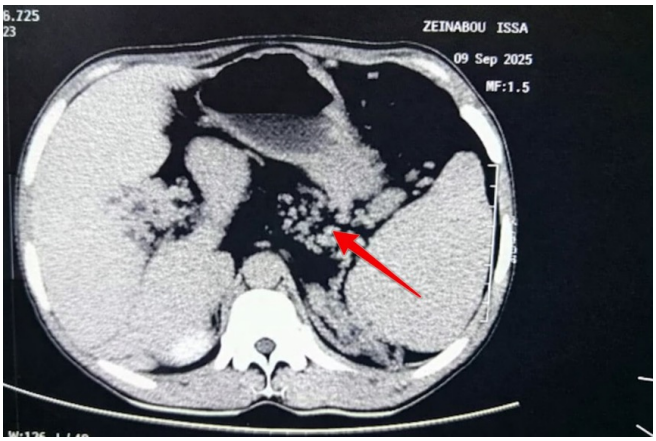

Figure 4. Axial abdominal CT scan showing a portal cavernoma with associated homogeneous splenomegaly.

Discussion

This is the first case of portal cavernoma reported at Zinder National Hospital. In Romania, Cazacu et al. found a prevalence of 14.8% among patients with portal thrombosis.[6] Despite its rarity, the condition is increasingly detected thanks to advanced imaging.

Our patient was 30 years old, within the range reported for non-cirrhotic portal vein thrombosis. In Senegal, Diallo et al. reported a mean age of 41 years (15–75 years), showing that the disease affects young adults. Although some series report a male predominance (M/F = 2.7: 1), gender differences may vary depending on underlying pro-thrombotic conditions sus as sickle cell disease.[5]

The causes of portal vein thrombosis are often multifactorial. Devika et al. in India identified local inflammatory factors (pancreatitis, abdominal infections such as diverticulitis) and systemic thrombophilia, such as hyperhomocysteinemia.[7] Resseguier et al. in France found in 24 of 32 cases, 15 haematological disorders (10 myeloproliferative syndromes, 2 antiphospholipid syndromes, 1 thalassaemia major, 1 hyperhomocysteinemia and 1 factor II mutation).[8] In children, umbilical vein catheterization is the main cause.[9] In our case, no aetiology was apparent, i.e. idiopathic (Elkrief et al.[3])

Upper gastrointestinal bleeding was the presenting symptom, as often described in the literature. Wei et al. highlighted variceal bleeding as the most common initial presentation.[10] Boccatonda et al. and Layton et al. also reported that variceal rupture is a typical manifestation of cavernous transformation.[1,2]

Splenomegaly is a classical sign of portal hypertension; its stage III presentation in our patient reflects advanced collateral circulation. Elkrief et al. emphasized that splenic hypertrophy is one of the most sensitive clinical indicators of chronic portal hypertension.[3] The presence of fever and malaria might have acted as a precipitating or aggravating factor for thrombosis.

Anaemia in our case was due to acute bleeding on chronic asymptomatic bleeding, while thrombocytopenia reflected hypersplenism secondary to portal hypertension. Cazacu et al. described similar haematological abnormalities, considering hypersplenism as a constant feature of cavernoma.[6] Normal liver function tests can help rule out cirrhosis.

Ultrasound scan remains the first-line diagnostic tool, especially in resource-limited settings. In our case, it showed serpiginous vascular structures in the

hepatic hilum. Colour Doppler revealed turbulent, multidirectional flow, characteristic of cavernous transformation. Marra et al. confirmed the high sensitivity of ultrasound for differentiating partial from complete portal thrombosis.[11] Kalra et al. emphasized that in chronic cases, the portal vein may disappear centrally and be replaced by a network of periportal collaterals.[12] A CT scan confirmed the diagnosis, showing multiple venous collaterals and splenomegaly without hepatic lesions. Mild biliary compression may occur, corresponding to portal biliopathy, a complication described in 5–30 % of cases.[13–14] Jha et al. reported that CT findings typically include non-enhancement of the portal vein, serpiginous collaterals, and absence of focal hepatic lesions in non-tumoral thrombosis.[15]

Endoscopy revealed grade III oesophageal varices. Boccatonda et al. also noted that large varices are common in cavernomas.[1] The absence of red signs suggests a lower immediate bleeding risk but requires secondary prophylaxis.

Management of portal cavernoma is challenging. Acute bleeding requires resuscitation, transfusion, and vasoactive or beta-blocker therapy after stabilization and correction of anaemia. We used propranolol for secondary prophylaxis. In chronic thrombosis, recanalization is often impossible, and the benefit of anticoagulation remains controversial. Boccatonda et al. argued against anticoagulation in fully organized thrombosis.[1] In well-equipped centres, endoscopic variceal ligation and trans-jugular intrahepatic portosystemic shunt (TIPS) are standard options for recurrent bleeding or refractory portal hypertension. However, these techniques were unavailable in our hospital, reflecting the therapeutic limitations in many African settings.

Prognosis depends on the control of variceal bleeding and the absence of complications such as portal biliopathy or intestinal ischaemia. Our patient showed favourable progress with improved haemoglobin and no recurrent bleeding after one month. Regular follow-up is essential to adjust beta-blocker dosage and consider further endoscopic evaluation.

Conclusion

Portal cavernoma represents the chronic evolution of portal vein thrombosis, often of unknown aetiology. Its discovery is usually prompted by complications of portal hypertension, such as variceal bleeding. Biological abnormalities are nonspecific, but imaging and endoscopy are key to diagnosis. Management must be multidisciplinary, combining resuscitation, beta-blockers, endoscopic treatment, and possibly TIPS or surgical shunts where available. In resource-limited environments, supportive care and close monitoring remain the cornerstone of management.

Conflict of interest: None

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