Reminder of important clinical lesson

Cavernous transformation of portal vein a missed cause of extrahepatic portal hypertension

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Summary

The portal vein thrombosis is an important and well-known cause of portal hypertension in young patients. Cavernous transformation of portal vein may be a sequel of portal vein thrombosis. Here the author reports a case of a 14-year-old boy presented with haemetemesis and melena, which was due to rupture of oesophageal varices as a result of portal hypertension. On further investigation, cavernous transformation of portal vein was identified on CT portovenogram and normal liver structure was evident on histology.

BACKGROUND

Extrahepatic portal vein obstruction is a common cause of portal hypertension in developing countries and constitutes up to 40% of all patients with portal hypertension. Cavernous transformation of the portal vein is an idiopathic angiomatous malformation. Cavernous transformation is an important cause of extrahepatic portal hypertension in children, besides the more common portal vein thrombosis (PVT).¹ The exact pathogenesis that leads to cavernous transformation has been disputed; some researchers suggest that it forms due to long-standing PVT,

in attempt to recanalise the portal vein, dilated multiple small vessels develop around the obstructed portal vein. Thrombosis and occlusion of the portal vein leads to portal hypertension with enlarged spleen and the development of portosystemic collaterals. Variceal bleeding is frequently the first sign of extrahepatic portal hypertension. Others may present with haematologic abnormalities due to hypersplenism. Here the author reports one such case of cavernous transformation of portal vein (CTPV). Also pathogenesis, clinical feature and management of CTPV are briefly discussed here.

Figure 1 The liver consists of a vast interanastomosing network of hepatocytes arranged in single-cell thick plates separated by vascular sinusoids.

Figure 2 Coronal CT portal venogram shows multiple collateral around the porta hepatis.

CASE PRESENTATION

A 14-year-old male was admitted in emergency unit for complaints of haematemesis and melena for 2 days. He had also been experiencing mild heaviness and dragging sensation in the left hypochondrium for last 2 months. The history was negative for jaundice, non-steroidal anti-inflammatory drug abuse, alcohol intake or any other addiction. There is no history of such illness in other family members. Clinical examination showed severe pallor, pulse 100/min, blood pressure 100/60 mm Hg and pedal oedema was absent. Abdominal examination revealed fullness of left hypochondrium in scaphoid abdomen but no visible veins. Spleen was palpable 5 cm below the left subcostal margin and firm. Liver was not palpable and rest of the examinations was normal.

INVESTIGATIONS

Laboratory findings were- haemoglobin-5.9 g/dl, total leucocyte count-1590/mm³ with differentials of polymorphs 60%, lymphocytes 34% and eosinophils 4% and platelet count 1.25 lacs/mm³, general blood picture was microcytic hypochromic. Blood sugars, electrolytes and kidney function tests were normal. Liver function tests showed total serum bilirubin 0.5 mg/dl, serum glutamic pyruvic transaminase 40 IU/l, serum glutamic oxaloacetic transaminase 49 IU/l, serum alkaline phosphatase 247 IU/l, serum protein 6.5 g/dl, serum albumin 4.2 g/dl, prothrombin time 15 s (normal upto 16 s) and international normalised ratio was 1.07. Viral markers for hepatitis B and C were negative. Ultrasonography abdomen revealed normal size and normal echotexture of liver, massive splenomegaly (16 cm) with portal vein diameter 11.3 mm and no free fluid in abdominal cavity. Upper gastrointestinal endoscopy revealed grade 3 oesophageal varices. Liver biopsy showed

normal morphology (figure 1). CT portal venogram (64-slice) was done which showed CTPV with multiple collaterals at the porta hepatis (figure 2).

DIFFERENTIAL DIAGNOSIS

- PVT
- Tropical splenomegaly, it is recognised cause of portal hypertension²
- Non-cirrhotic portal fibrosis
- Congenital hepatic fibrosis (CHF)
- Schistosomiasis.

TREATMENT

The patient was managed conservatively with intravenous fluid, octreotide, proton pump inhibitors and two units of whole blood were transfused.

OUTCOME AND FOLLOW-UP

After being stabilised the patient was discharged on β blocker, along with proton pump inhibitors and haematinics. The patient was referred for portosystemic anastomosis.

DISCUSSION

CTPV is a relatively rare condition found in about 3.2% cases of portal hypertension.³ CTPV is an uncommon cause of portal hypertension in adults, Some researches consider this condition to be congenital, while others accept that it is a consequence of PVT due to neonatal umbilical vein sepsis or a complication of an inflammatory, neoplastic process occurring later in life.⁴ Inherited and acquired disorders of coagulation pathway may also be causative. Inherited disorders include mutations in the

prothrombin gene G20210A as well as deficiencies of various intrinsic anticoagulation factors, such as protein C and protein S, and activated protein C resistance. Acquired disorders include antithrombin III deficiency resulting from malnutrition, sepsis, disseminated intravascular coagulation, inflammatory bowel disease, liver disease or estrogen use.

CHF is an autosomal recessive disorder that belongs to the family of fibropolycystic liver diseases. Clinically fibropolycystic diseases have three effects being present in different proportions, those of a space occupying lesion, of portal hypertension and of cholangitis. CTPV has been found in association with CHF in about 48% cases.^{3 5}

Portal vein obstruction does not affect liver function unless the patient has an underlying liver disease such as cirrhosis.⁶ This is partially due to a rapid arterial buffer response, with compensatory increased flow of the hepatic artery maintaining the total hepatic blood flow. The collaterals may develop as early as 12 days after thrombosis, though the average time to formation is approximately 5 weeks.

Rarely, the thrombosis extends from the portal vein to the mesenteric arcades, leading to bowel ischaemia and infarction. Typically, patients with CTPV do not have cirrhosis and maintain normal hepatic lobular architecture with normal hepatic function. Onset of symptoms and signs is highly variable and ranges from early childhood to the fifth or sixth decades of life, although this disorder is diagnosed in most patients during adolescence or young adulthood.⁵

The therapy of CTPV is no more different from treatment of portal hypertension; it involves the prophylaxis of the first bleeding (primary prophylaxis), treatment of the bleeding episode and prevention of a recurrence of

variceal bleeding (secondary prophylaxis). The pharmacotherapy, endoscopic therapy and surgical intervention are the methods used for prophylaxis of variceal bleeding and rebleeding.

The clinical diagnosis of extrahepatic portal venous obstruction was confirmed by portal venography in our case and also corroborated by normal liver histology.

Learning points

- ▶ CTPV is an important and often overlooked cause of extrahepatic portal hypertension in children.
- ▶ In our case, 64-slice CT scan has enabled us to diagnose CTPV and has recently become a safe and valuable technique for evaluating such cases.

Competing interests None.

Patient consent Obtained.

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