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SHORT REPORT

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Hepatoportal sclerosis in a child

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Hepatoportal sclerosis (non-cirrhotic portal fibrosis) is characterised by portal hypertension, splenomegaly and variceal bleeding. It is rarely seen in childhood. We report on hepatoportal sclerosis in a patient with haematemesis as a presenting symptom.

This 11-year-old male patient was admitted with upper gastrointestinal bleeding (UGB). He was observed for UGB for the first time at the age of 6 years at another hospital and was treated with sclerotherapy. Afterwards he had no bleeding from varices until admission to our hospital. At the age of 6 months, he had been admitted and treated with antibiotics and intravenous fluid because of diarrhoea.

On physical examination, body weight and height were at the 10th–25th percentile, the spleen was enlarged (7 cm under the costal margin) but the liver was not palpable. There was no collateral circulation on the abdominal skin. On Doppler ultrasonography, the liver parenchyma was coarsely granular, periportal hyperechogenicity was detected, and the inferior vena cava and hepatic veins were patent. At the portal hilus, a vascular structure resembling cavernous transformation or a recanalised portal vein was seen. The portal vein diameter was enlarged with multiple collaterals. Haemoglobin was 10.1 g/dl, leukocytes 4100/mm^3, thrombocytes 145000/mm^3, prothrombin time 18 s (reference range 11–15 s), activated partial thromboplastin time 35 s (reference range 25–35 s), international normalised ratio (INR) 1.5, blood glucose, renal function (BUN 10 mg/dl, creatinine 0.6 mg/dl) and electrolytes (sodium 139 mmol/l, potassium 3.8 mmol/l, chloride 102 mmol/l) were normal. Aspartate and alanine aminotransferase levels were 43 and 34 U/l, respectively. Total bilirubin level was 0.4 mg/dl. Serum albumin level was 3.8 g/dl, gamma glutamyltranspeptidase and alkaline phosphatase activity were 56 and 480 U/l, respectively.

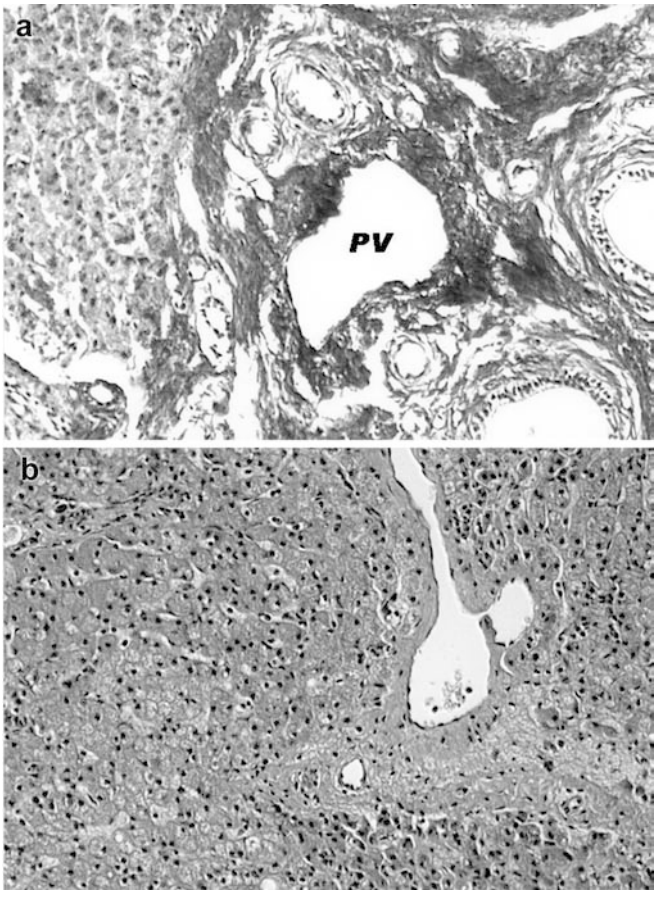

Fig. 1 **a** Subendothelial fibrosis of the portal vein (*PV*) branch with an irregular luminal contour (Elastic von Geisson ×100). **b** Thickening in the wall of portal vein branch with herniation of the vessel into the hepatic parenchyma (H and E ×100)

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Hepatotropic viruses, urine and blood amino acids, serum copper, coeruloplasmin, iron, transferrin saturation, ferritin, alpha 1-antitrypsin, sweat chloride test, protein C and S levels were normal. Anti-smooth muscle, anti-liver-kidney microsomal type-1 and anti-nuclear antibodies were negative. On upper gastrointestinal endoscopy, there were three columns of varices at the oesophagus and were treated by sclerotherapy. A percutaneous liver biopsy was normal. On abdominal laparotomy, the portal system was patent and a cavernous transformation was not seen. A wedge liver biopsy was taken. Histopathological examination revealed a preserved liver architecture. The portal tracts, hepatic arteries and bile ducts appeared normal. Fibrous bands were present extending from internal capsule to the parenchyma and from portal areas to the adjacent parenchyma. Portal vein branches showed subendothelial fibrous thickening resulting in a thickening of the vessel wall and an irregular luminal contour (Fig. 1a). Additionally, aberrant portal vein branches protruding from portal tract borders were seen (Fig. 1b). No thrombi were detected. There was no inflammatory infiltrate. Propranolol was started. After 1 month, two columns of oesophageal varices were seen on endoscopic examination and sclerotherapy was performed. During a 13-month follow-up period following the second sclerotherapy session, haematemesis or melaena did not recur.

Hepatoportal sclerosis, mainly seen in adults, is a rare cause of portal hypertension in childhood. The aetiopathogenesis of this disease is not understood. Exposure to chemical agents such as vinyl chloride, copper sulphate, azathioprine and arsenic has been incriminated [5, 7, 10]. Intestinal bacterial infections with repeated septic embolisation of the portal circulation have been proposed as a possible aetiology [9]. The latter hypothesis is supported by the fact that the disease is frequent in developing countries [9]. Umbilical sepsis in newborns, bacterial infections and diarrhoeal episodes in infants and in early childhood are likely to lead to portal pyaemia and pylophlebitis, resulting in thrombosis, sclerosis and obstruction of small- and medium-sized portal vein radicals [9]. Our patient had a history of diarrhoea at 6 months of age.

Patients with hepatoportal sclerosis are generally admitted with UGB and abdominal discomfort due to splenomegaly [3, 8, 9]. As a rule, there is no massive hepatomegaly in these patients [8, 9]. Hepatic functions are usually preserved [9]. A small group of patients with ascites and hypoalbuminaemia has been noted to have nodular transformation of the liver with extensive portal and sub-hepatic fibrosis [3]. In our patient, the albumin level was low and nodular formation and extensive fibrosis were seen at biopsy. Prognosis of these patients is generally good, but occasionally fatal bleeding may occur.

In a recent paper, INR in non-cirrhotic portal fibrosis patients was reported to be significantly prolonged (1.8) as compared to healthy controls (mean INR 1.3) [1]. Prothrombin time and INR were also prolonged in our patient. The coagulation defects seen in these patients are suggestive of a sub-clinical hepatic involvement affecting the liver-dependent clotting factors, resulting in the prolongation of the INR without greatly affecting the partial thromboplastin time [1].

A percutaneous liver biopsy may fail to detect subcapsular fibrosis and vascular changes, so that the diagnosis may be overlooked as in our patient [6]. In these patients, a wedge liver biopsy may be necessary [3, 6]. In the pathological assessment of biopsy specimens of such patients, the main histopathological change is sclerosis or obliteration of portal vein branches resulting in formation of aberrant portal veins in portal tracts [6]. Septa extending from portal areas towards neighbouring tissues, pseudonodule formation, and extension of fibrosis into the hepatic parenchyma from subcapsular areas may be seen [3]. Most of these findings were observed in our patient's biopsy specimen. Piecemeal or hepatocyte necrosis and regeneration and vasculitis or active thrombosis were not observed in our patient.

Endoscopic sclerotherapy is the treatment of choice. Sclerotherapy (six times on average (range 1–14) for a patient) was effective enough to obliterate varices and 95% of UGB episodes were controlled by sclerotherapy [2, 9]. Prophylactic propranolol administration may also be used [6]. Liver transplantation is necessary in patients with hepatocellular deficiency and complications [4].

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