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Case report

Severe gastrointestinal tract bleeding in a two-month-old infant due to congenital intrahepatic arterioportal fistula

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Abstract

A 2-month-old boy was referred for assessment of severe upper gastrointestinal tract bleeding and melena. On physical examination, a continuous murmur was heard over the right upper quadrant of the abdomen. A splenomegaly and dilated veins were also noted on the abdominal wall. Liver functions were normal. There was no history of trauma or jaundice. Doppler ultrasonography, magnetic resonance arteriography and angiography suggested the presence of an intrahepatic arteriovenous fistula between the phrenic artery and the portal vein.

Management consisted of successful embolization by coiling of the phrenic artery.

To our knowledge this is the first documented case report of a congenital fistula between the phrenic artery and the portal vein.

Keywords: Congenital; Arterioportal fistula; Gastrointestinal bleeding; Embolization

1. Introduction

Congenital arterioportal fistulas (APF) are rare, being mostly described after trauma or in the case of a tumor (haemangio endothelioma, hepatoblastoma and angiosarcoma) [1]. The clinical recognition is important because life threatening bleeding due to severe portal hypertension may occur. The clinical manifestations are complications of portal hypertension, most commonly bleeding from varices in the gastrointestinal tract [2,3]. On physical examination, a continuous murmur over the right quadrant of the abdomen, a splenomegaly and dilated veins on the abdominal wall may be present. Ultrasound is the first performed imaging modality [4]. Arteriography confirms the diagnosis, which is often followed by the definitive therapy of coiling of the artery.

We describe the case of a 2-month-old boy who was referred for assessment for upper gastrointestinal tract bleeding and melena. Ultrasound, magnetic resonance angiography (MRA) and arteriography revealed an intrahepatic arterioportal fistula.

2. Case report

A 2-month-old boy was referred to our hospital presenting with upper gastrointestinal tract bleeding and melena. Medical history revealed no abdominal trauma or hepatic disease. The baby was breast-fed and received additional Vitamin-k supplement as recommended by the current Dutch guidelines [5]. On physical examination, the patient was pale and dystrophic. He weighed 4135 g (−2 SD). Blood pressure was 83/44 mmHg and a pulse rate of 160 beats/min. The abdomen was extended and tender at palpation. The liver was 1–2 cm and the spleen 4 cm palpable below the right and left costal margin, respectively. A continuous murmur was heard over the right upper quadrant of the

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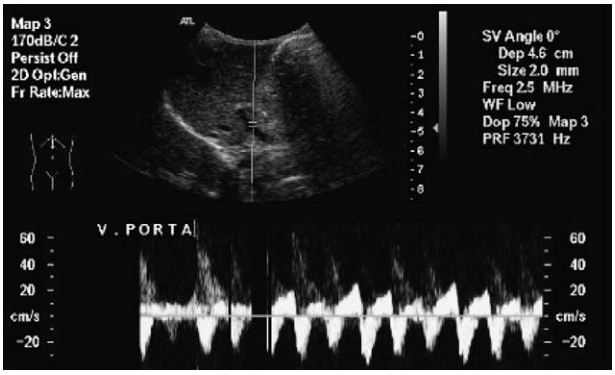

Fig. 1. Ultrasound of the liver with the doppler in the main portal vein. The spectrum shows an arterialisation of the signal.

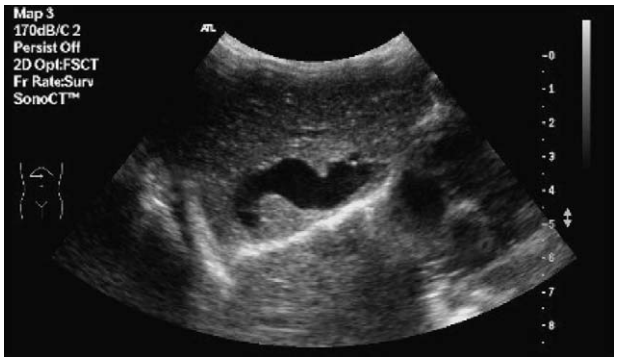

Fig. 3. Ultrasound of the liver. Transverse image which shows a widely dilated intrahepatic portal vein.

abdomen. Dilated veins were present on the abdominal wall.

The patient's hemoglobin (Hb) level was below normal at 4.0 mmol/l (5.9–8.4); thrombocytes count was 338×10^9/l (150–350). Factors involved in the synthetic function of the liver were normal.

Prothrombin time (PT) and activated partial thromboplastin time (APTT) were 23 and 33 s, respectively. The values of the fibrinogen and D-dimers were 910 mg/l (1700–4000) and 3640 ng/ml, respectively (normal values: fibrinogen 2 gm/l; D-dimer, 500 ngm/ml).

An erythrocyte transfusion was given to restore normal Hb levels. Because of the extended and tender abdomen with a continuous murmur, a Doppler ultrasonography (US) was performed. It showed a homogeneous echo pattern of the liver. The gallbladder and bile ducts were of normal aspect and size. There was a slight hepatomegaly. In the right quadrant of the liver an intrahepatic widely dilated portal vein was noted, which was also found to be connected with an artery running on top of the liver. The portal and lienalis veins were dilated with a hepatofugal flow. Doppler US showed a turbulent flow with arterial spikes in these veins. In the parenchyma, surrounding the portal veins, color artifacts were visible. The inferior caval vein demonstrated a normal size and there was no dilatation of the hepatic veins. The size of the spleen was 7 cm. There was a small amount of ascites. A diagnosis of an arterioportal fistula was made (Figs. 1–3). To confirm the diagnosis a MRI was performed. Unfortunately due to movement and respiratory artifacts, no additional information was obtained.

Subsequently an angiogram was performed. The selective angiogram of the celiac and superior mesenteric arteries was normal. However, just lateral of the origin of the superior mesenteric artery, an artery was visualized originating from the aorta connecting with the right portal vein, filling and dilating the superior mesenteric and lienalis veins (Fig. 4a and b). Coils (VortX Diamond Shape Occlusion Coils®, Boston Scientific, USA, 2×5 mm and 1×3 mm; Microcoil soft platinum, Cook, Denmark, 1×7 mm) were successfully inserted into the phrenic-portal fistula with a total occlusion of the

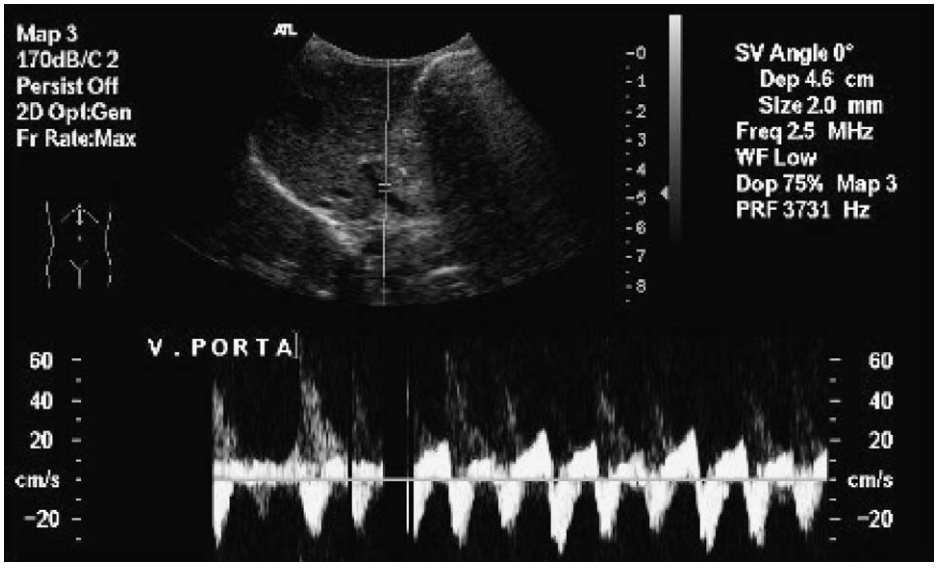

Fig. 2. Ultrasound of the abdomen in the midline. The doppler is positioned in the proximal part of the splenic vein. The spectrum shows an arterial signal.