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Case report

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

R. Aarts [a], M.M. Ijland [b], I. de Blaauw [c], Y. Hoogeveen [a], C. Boetes [a,*], M. van Proosdij [a,1]

[a] *Department of Radiology, University Medical Center St. Radboud, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands*
[b] *Department of Pediatrics, University Medical Center St. Radboud, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands*
[c] *Department of Pediatric Surgery, University Medical Center St. Radboud, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands*

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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

* Corresponding author.
E-mail address: C.Boetes@rad.umcn.nl (C. Boetes).
[1] Present address: Medisch Centrum Alkmaar, Postbus 501, 1800 AM Alkmaar, The Netherlands.

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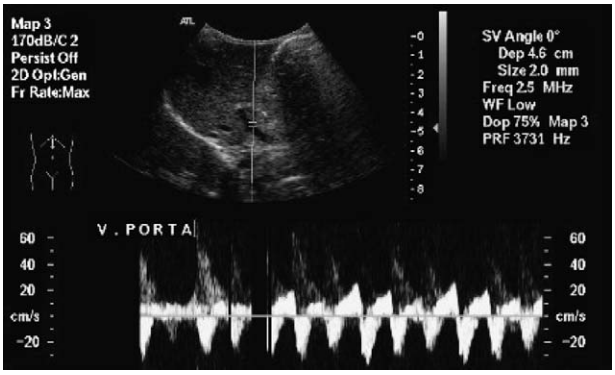


Fig. 1. Ultrasound of the liver with the doppler in the main portal vein. The spectrum shows an arterialisation of the signal.

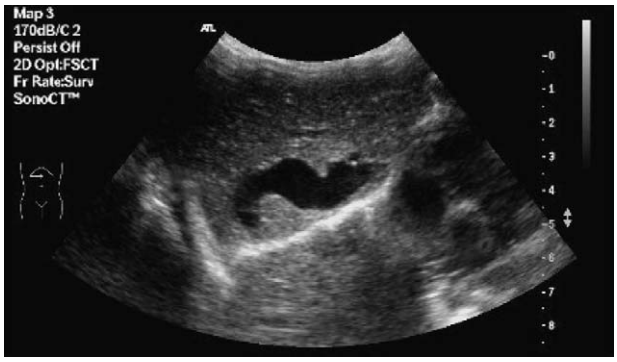


Fig. 3. Ultrasound of the liver. Transverse image which shows a widely dilated intrahepatic portal vein.

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

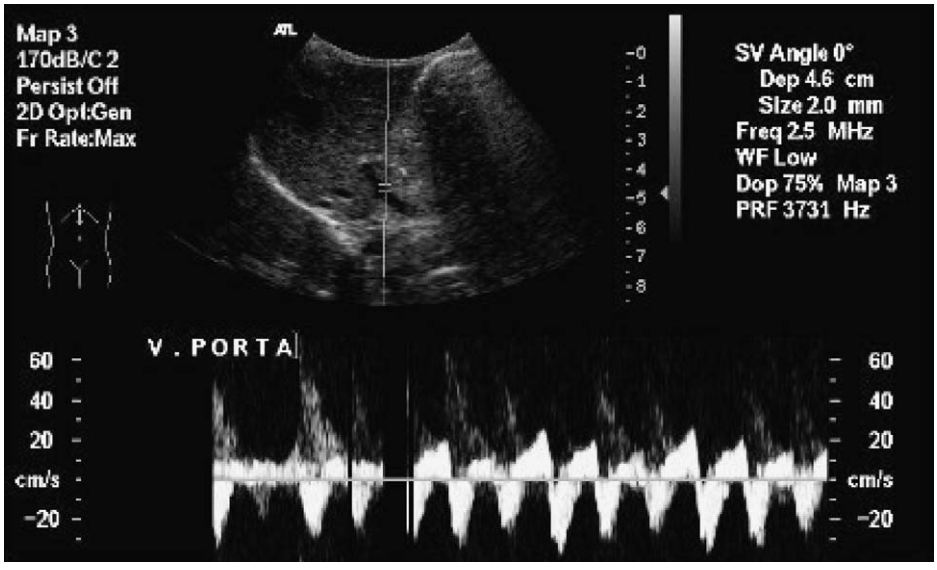
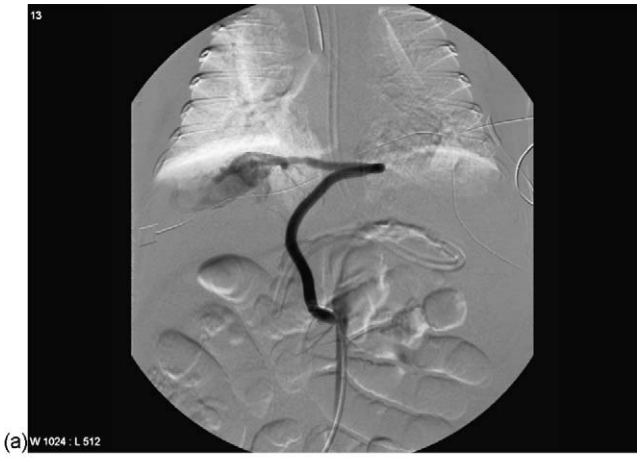


Fig. 2. Ultrasound of the abdomen in the midline. The doppler is positioned in the proximal part of the splenic vein. The spectrum shows an arterial signal.



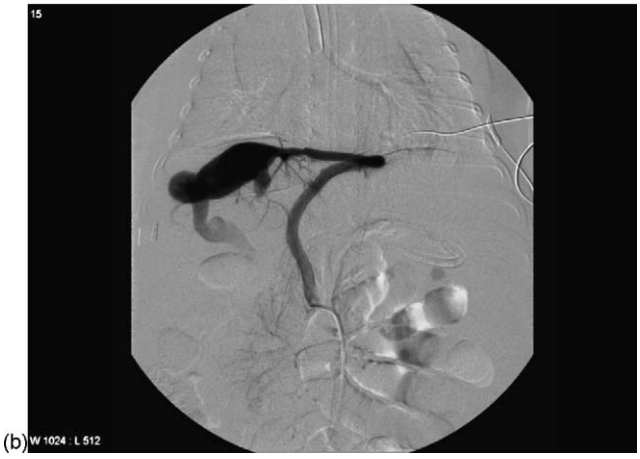


Fig. 4. (a and b) Selective angiogram with a four French Cobra catheter positioned in the proximal part of the phrenic artery. The image shows a connection with a widely dilated portal vein (see also ultrasound image).

fistula and normal filling in the late phase of the portal and superior mesenteric vein were achieved (Figs. 5 and 6). There were no complications during or after the procedure. Four days after the intervention ultrasound demonstrated a normal

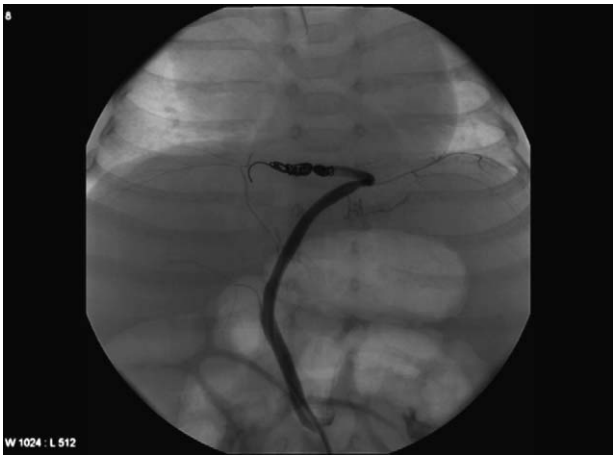


Fig. 5. Angiogram after occlusion of the fistula with microcoils. The image shows an absent filling of the fistula.

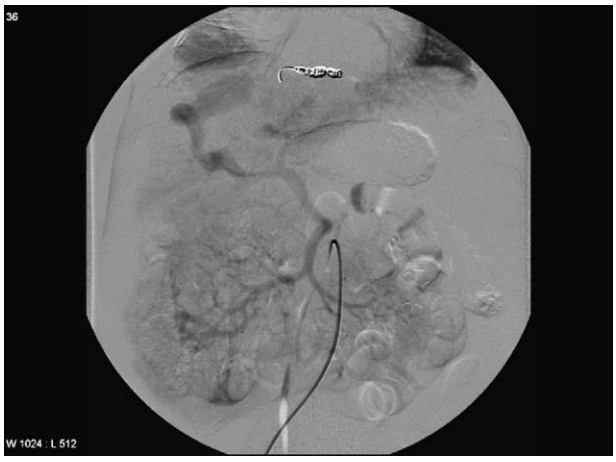


Fig. 6. Angiogram of the mesenteric artery after occlusion of the fistula shows a normal filling of the main portal vein and the intra hepatic portal vein branches including the widely dilated vessel.

venous signal and flow in the portal vein. An upper gastrointestinal endoscopy showed no varices.

At 3 months follow-up physical examination and routine laboratory workup were found to be normal. The hepatosplenomegaly and hepatic murmur were no longer present. His postoperative weight at the ages of 5 months was 6240 g. At abdominal ultrasound, flow in the right portal vein was observed to be absent. The portal vein was not dilated and demonstrated normal hepatoportal flow without arterialisation of the signal.

3. Discussion

We describe a congenital APF between the phrenic artery and the portal vein, which to our knowledge is the first documented case.

Almost two-thirds of APF originate from the hepatic artery. The superior mesenteric artery and the splenic artery are other locations [2]. The phrenic artery is a branch of the abdominal aorta and ascends towards the diaphragm. Its course resembles the course of the artery in our case. We did not find any data concerning congenital arterioportal fistula between the phrenic artery and the portal vein. An APF is a relatively rare acquired or congenital disorder. Acquired APF are mostly described after blunt or penetrating trauma, iatrogenic procedures, tumors and aneurysms [2,6–9]. Congenital APF are associated with hereditary telangiectatic diseases (Ehlers-Danlos syndrome, Osler-Weber-Rendu syndrome), arteriovenous malformations, biliary atresia and aneurysms [2,4,10–12]. Vauthey et al. [2] reported that congenital causes are responsible for about 15% of the cases.

Arterioportal fistulae can be asymptomatic but often present with complications of portal hypertension, including lower or upper gastrointestinal bleeding, ascites, diarrhea, hepatosplenomegaly or heart failure [2,3]. Additional clinical features are a bruit or a thrill over the right quadrant

of the abdomen and hepatosplenomegaly. Failure to thrive is also a common presentation in infants, due to intestinal dysfunction which results in malabsorption, diarrhea and steatorrhea [8,9]. The severity of symptoms is likely to be related to the amount of blood shunted, the location of the fistula and the resistance of the liver to the increased portal venous flow [13,14]. For our patient, upper gastrointestinal bleeding, probably originating from esophageal varices and melena, were the first presenting symptoms. When gastrointestinal tract bleeding presents in such a young baby one should also consider bleeding due to bacterial enteritis, milk protein allergy, intussusception and anal fissure. Rare causes include volvulus, Meckel diverticulum, stomach ulcer and coagulation disorders [15]. In our patient the specific symptoms of a continuous murmur over the right upper quadrant of the abdomen, the dilated abdominal veins and the slightly enlarged liver and spleen were an indication to perform a Doppler US, which ultimately led to the diagnosis. Doppler US is the first imaging modality of choice. Common findings include enlargement of the hepatic artery and dilatation of that segment of the portal vein where the fistula is located. Doppler US features of congenital arterioportal fistula include pulsatile hepatofugal flow in the portal vein and color speckling in the hepatic parenchyma adjacent to the fistula (vibration artifact) [4]. Perfusion anomalies of the adjacent parenchyma can also be demonstrated, such as regional increase in arterial inflow as a response to the inverted portal flow, increase in portal vein inflow due to the shunt itself [4,10,16]. If left untreated, arterialisation of the portal vein causes the early onset of portal hypertension. Hepatoportal sclerosis and fibrosis of the portal radicles subsequently develop, further contributing to portal hypertension. Embolization of the feeding artery with or without subsequent surgery is currently the preferred therapeutic option. It is important to closely monitor affected patients because the fistula may recur through arterial collateralisation [17,18].

In conclusion, gastrointestinal bleeding due to portal hypertension may, in rare instances, be caused by arterioportal fistulas. It is important to recognize arterioportal fistulas early as associated complications are serious. Selective arteriography can confirm the diagnosis and often affords the definitive therapy in the same session.

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