Definitive therapy for a rare cause of recurrent ascites

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Summary

Extrahepatic arterioportal fistulas are a rare cause of portal hypertension in infants and adults. Most cases of arterioportal fistulas in adults are due to trauma, liver biopsy, surgery or tumours. The authors present a case of arterioportal fistula diagnosed in adult life without any of the above aetiologies. The patient became acutely symptomatic with diarrhoea and ascites at the age of 66 years and was successfully treated with a single endovascular embolisation procedure after failed medical therapy.

Introduction

The most common causes of recurrent ascites are liver cirrhosis, heart failure and peritoneal malignant disease. In these contexts, the management of recurrent ascites is challenging with significant side effects resulting from the treatments. We present a case of a rare cause of recurrent ascites with a single definitive radiological treatment.

Arterioportal fistulas (APFs) cause arterialisation and increased pressure in the portal venous system due to an anomalous connection from upper gastrointestinal arteries, usually hepatic arterial branches, to the portal venous system. The APF can be intra- or extrahepatic. The fistula is often a single connection, but may involve multiple branches of the arterial and venous systems.^{1–3} These anomalous connections have most commonly been observed to originate from the hepatic artery (65%), splenic artery (11%) and superior mesenteric artery (10%).⁴

When APFs become symptomatic in adults, they most commonly present with features of portal hypertension. The most common clinical presentations are gastrointestinal bleeding (33%) and ascites (26%), with other symptoms including diarrhoea, hepatic encephalopathy and heart failure.³ A bruit, or thrill, is a valuable clinical finding in diagnosing APFs.

Although standard single-phase CT can identify the presence of APFs, they are best diagnosed with both arterial and portal venous phases through the liver.

The technique of radiological endovascular embolisation has been successfully used for closure of APFs in paediatric¹ and adult patients. Repeat procedures are occasionally required.⁵ We are aware of one death following such a treatment in an adult when occlusion of the APF led to progressive intrahepatic cholangitis and, ultimately, death.⁶

Case report

A 66-year-old man was referred urgently by his general practitioner to the gastroenterology outpatient clinic for assessment of acute onset diarrhoea for 6 weeks and increased abdominal girth over 2 weeks. The frequency of diarrhoea was six motions a day with no rectal bleeding and Bristol Stool Chart grade 5 consistency.⁷

His medical history consisted of sigmoid colectomy for diverticular disease 33 years previously, diabetes mellitus for 27 years, hypertension and ischemic heart disease with coronary artery bypass surgery 4 years previously. He had no history of liver disease, nor relevant family history. In addition, there was no history of liver biopsies or trauma. His recent alcohol history consisted of 2–4 units per month and rarely above 10 units a week previously.

Drugs history consisted of Novomix³⁰ 40 units twice daily, Simvastatin 40 mg at night, and Ramipril 5 mg, Atenolol 50 mg, Aspirin 75 mg, Lansoprazole 30 mg, Doxazosin 2 mg, Furosemide 20 mg and Codeine 30 mg in the morning.

On examination, no abnormalities were found in the cardiorespiratory system. Abdominal examination revealed a distended tight abdomen with shifting dullness consistent with significant ascites and

Figure 1 Arterial-phase CT scan showing contrast in the portal vein (i).

Figure 2 Angiogram via the replaced hepatic artery demonstrating a large arteriportal fistula with rapid retrograde filling of a dilated portal vein.

Figure 3 The post-embolisation angiogram demonstrating complete occlusion of the fistula with preservation of hepatic arterial flow.

generalised discomfort on palpation without peritonism. There were no signs of chronic liver disease or lymphadenopathy. His vital signs were all within normal range. He weighed 66 kg, with a body mass index (BMI) of 23.4 kg/m², having weighed 89 kg (BMI 31.5 kg/m²) in 2005.

Initial serology tests showed a high alkaline phosphatase of 804 IU/l (100–335 IU/l), raised γ -glutamyl transpeptidase of 303 IU/l (<60 IU/l) with otherwise normal liver function tests. His clotting showed a mildly prolonged prothombin time representing an international normalised ratio of 1.4. He had normocytic anaemia with haemoglobin of 9.4 g/dl (13.5–17.5 g/dl) and his platelet count was within the normal range. Of note, the serological liver screen was negative including viral, autoimmune and metabolic indices. Stool microscopy and culture were also negative.

Abdominal ultrasound confirmed the presence of gross ascites with normal liver size and echogenicity, with a mildly enlarged spleen. Ascitic fluid analysis showed a white cell count of 0.3×10^9 /l and serum ascites albumin gradient of 19 g/l denoting portal hypertension. An upper gastrointestinal endoscopy revealed mild gastric inflammation with no evidence of varices.

Single-phase CT of the abdomen suggested the presence of abnormal vessels within the right lobe of the liver. This was confirmed on a subsequent

dual-phase CT (figure 1), which showed an enlarged replaced hepatic artery arising from the proximal superior mesenteric artery together with arterialisation of the portal venous system, diagnostic of an APF. In addition, the right portal vein branch was markedly dilated due to increased pressure from arterialisation.

A CT-guided transcutaneous liver biopsy was also performed which showed macronodular cirrhosis with portal veins demonstrating intimal smooth muscle proliferation consistent with arterialisation of the liver. There was no interface hepatitis, inflammation, steatohepatitis or iron retention and no other aetiological clues.

The patient was initially treated with a salt-restricted diet and addition of Spironolactone up to 300 mg per day, but due to poor response to medical therapy an abdominal paracentesis was performed which drained 6 l of ascites. While on increasing diuretic therapy a further re-accumulation of ascites within 4 weeks necessitated a further 10 l paracentesis.

Due to the rapid onset, recurrent nature of the ascites and failed medical therapy, the APF was definitively treated with endovascular embolisation 3 months after presentation, and subsequent successful weaning off all additional diuretics.

Embolisation was achieved through bilateral femoral arterial access and the initial angiogram (figure 2) via

the replaced hepatic artery demonstrated a large APF with rapid retrograde filling of a dilated portal vein. The right hepatic arterial branch feeding the fistula was occluded using platinum coils. The post-embolisation angiogram (figure 3) demonstrated complete occlusion of the fistula with preservation of hepatic arterial flow.

All features of portal hypertension were resolved with this single procedure and a CT scan performed 3 months post-embolisation demonstrated persisting occlusion of the APF and no filling of the portal venous system in arterial phase. The patient continues to be asymptomatic at 1-year follow-up.

Discussion

When APFs develop in adults, they are almost exclusively secondary to abdominal trauma, liver biopsy, surgery or tumours. In this report, we describe a case without the presence of any of these recognised aetiological factors. The sigmoid colectomy 33 years previously may have been the triggering event, but the prolonged timescale does not offer a satisfying cause for the creation of the APF. Rarer causes of APFs have been reported, such as ruptured pancreatic pseudocysts and superior mesenteric aneurysms.^{8,9} No evidence of either of these aetiologies was present in this case, but they do demonstrate that other causes for this rare condition are still being discovered.

The presence of cirrhosis in the liver biopsy in this case is also worth considering as a cause or effect of the APF. The causative relationship between cirrhosis and APFs is unclear. In canine studies, large APFs caused focal fibrosis in the periportal zones, periportal necrotising vasculitis and increased hemosiderin deposits.⁸ These features were not present in this case. An external experienced liver pathologist reviewed the biopsy and considered that given the presence of portal vein intimal smooth muscle proliferation consistent with hepatic arterialisiation of the liver, and paucity of other diagnostic features, the cirrhosis was more likely to have been caused by the APF, although the opposite scenario could not be excluded. In light of the history of diabetes, hypertension and historic obesity, non-alcoholic fatty liver disease was considered, but the biopsy findings did not support this as a cause of the cirrhosis. Furthermore, the patient had undergone ultrasound examination of the abdomen 3 years previously for potential gallstone disease, revealing a normal-sized liver, outline and echogenicity, with no evidence of portal hypertension or splenomegaly. His platelets were within the normal range up to and including this admission.

Guzman and colleagues have proposed a novel classification for APFs, with therapeutic implications.⁴ Small intrahepatic fistula caused by liver needle injury are classified as type I, with a recommendation for 1-month follow-up with Doppler and embolisation in those that persist past 1 month, or earlier, if haemodynamically significant. Type II APFs are larger intra/extrahepatic APFs with elevated portal pressures. It is recommended that all such APFs be treated by embolisation if technically possible or surgery if this is not feasible or unsuccessful. Finally, for type III APFs, which are congenital and are usually intrahepatic, the authors recommend referral to a specialised paediatric hepatobiliary centre. Treatment options include hepatic artery ligation, embolisation, resection or liver transplant. In this case, the aetiology of the APF is unknown but would satisfy classification as type II.

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