CASE REPORT

Portal biliary ductopathy caused by cavernous transformation of the portal vein

Jeffrey Chu-Nan Chong, Ray Boyapati, Ferry Rusli

Department of Gastroenterology,
Southern Health, Clayton,
Victoria, Australia

Correspondence to

Dr Jeffrey Chu-Nan Chong,
chongster.rocks@gmail.com,
jeffrey.chong@southernhealth.
org.au

SUMMARY

A 42-year-old woman presented with a 1-week history of epigastric pain and deranged liver function tests (LFTs) on a background of known portal vein thrombosis (PVT) with cavernous transformation. Imaging with ultrasound, CT and MR cholangiopancreatography demonstrated known PVT, with distortion of the common bile duct and a bulky head/proximal body of the pancreas, thought to be due to mass effect from cavernous transformation related to PVT. At endoscopic retrograde cholangiopancreatography, the common hepatic duct was noted to be smaller in diameter, without any filling defects or discrete strictures. Sphincterotomy and balloon trawl was performed, with subsequent improvement of the woman's LFTs and abdominal pain.

BACKGROUND

Cavernous transformation of the portal vein is a rare condition, with various aetiologies and diverse clinical presentations. It is known to be a long-term sequelae of portal vein thrombosis (PVT).¹ Complications are associated with portal hypertension and most published literature focuses on diagnosis and management of portal hypertension in this setting.^{2–5} Symptomatic biliary obstruction caused by cavernous transformation of the portal vein is an extremely rare disorder for which no consensus to optimal treatment exists.⁶ Endoscopic management of biliary strictures caused by cavernous transformation of the portal vein appears to be effective and safe.⁶ In the event of recurrent failure of endoscopic treatment, surgical management and ultimately, liver transplant has been performed.^{4 7–9} This case demonstrates safe endoscopic management of a presumed biliary tree obstruction on a background of portal biliary ductopathy due to cavernous transformation of the portal vein.

CASE PRESENTATION

A 42-year-old woman presented to the hospital with a 1-week history of severe epigastric pain. The pain was described as constant, sharp and with no radiation. It was associated with one episode of vomiting. She was found to have deranged cholestatic liver function tests (LFTs). On examination, she had a soft abdomen, with epigastric tenderness.

Her medical history consisted of a spontaneous PVT with cavernous transformation. She had previously been on warfarin but on admission, was not on any anticoagulation. She also had type 2 diabetes mellitus and had previously had a splenectomy for hereditary spherocytosis.

INVESTIGATIONS

Ultrasound of the abdomen demonstrated known PVT, with no flow in the right or left portal veins. MR cholangiopancreatography demonstrated splaying of the right and left hepatic ducts, with posterior displacement/distortion of the common bile duct (CBD) and a bulky head/proximal body of the pancreas. The posterior displacement of the CBD at the level of the porta hepatic was thought to be due to mass effect from cavernous transformation related to PVT (figures 1 and 2). CT of the abdomen confirmed these findings and demonstrated a diffusely bulky head of pancreas.

On admission, alkaline phosphatase (ALP) was 205 U/l (normal <120 U/l), γ -glutamyl transpeptidase (GGT) 181 U/l (normal <64), alanine transaminase (ALT) 87 U/l (normal <56) and bilirubin 40 U/l (normal <20). LFTs continued to rise and reached a peak 1 week postadmission with an ALP of 366 U/l, GGT 207 U/l, ALT 105 U/l and bilirubin 60 U/l.

LFTs 1 month prior to admission demonstrated mildly abnormal ALP and GGT (133 and 99 U/l, respectively). ALT and bilirubin were normal at this time.

DIFFERENTIAL DIAGNOSIS

The major differential diagnoses in this case are chronic cholestasis due to portal biliary ductopathy or obstruction secondary to cholelithiasis. It can be difficult to distinguish between the diagnoses, as both can coexist. Cholestasis, one of the main clinical features of portal biliary ductopathy, is thought

Figure 1 MRI series 10, image 16: fat saturated T2-weighted images. Large calibre bile duct narrowing down to low calibre, adjacent to large heterogenous mass, which is consistent with blood of various ages, probably portal vein thrombus.

To cite: Chong JC-N, Boyapati R, Rusli F. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008339

Figure 2 MRI series 10, image 21: fat saturated T2-weighted images. Large calibre bile duct narrowing down to low calibre, adjacent to large heterogenous mass, which is consistent with blood of various ages, probably portal vein thrombus.

to result from the mass effect of enlarged collaterals or chronic thrombus compressing the biliary lumen. However, although biliary abnormalities are associated with PVT, the majority of cases remain asymptomatic.¹⁰

Cholelithiasis is also associated with PVT and portal biliary ductopathy, probably resulting from a combination of factors including the presence of strictures, loss of biliary motility and chronic cholestasis. Incidence and prevalence of cholelithiasis in this setting is difficult to determine due to lack of uniformity in investigation methods across a range of studies.¹¹

TREATMENT

At endoscopic retrograde cholangiopancreatography (ERCP), performed during admission, a sphincterotomy was performed. Cholangiogram demonstrated a symmetrically narrowed common hepatic duct that drained well (figure 3). A 10–12 mm balloon was dragged through the system without resistance.

Figure 3 Endoscopic retrograde cholangiopancreatography: demonstrates biliary tract narrowing at cholangiogram. Narrowing persists with time.

OUTCOME AND FOLLOW-UP

Following ERCP, the patient's epigastric pain improved significantly, as did her LFTs. She was discharged home 3 days post-procedure. It is possible that a small stone was removed during ERCP to account for improvement in her symptoms. She continues to be followed up by the haematology clinic.

DISCUSSION

In a review of portal hypertension due to portal venous thrombosis, Harmanci *et al*¹¹ suggested that the presence of portal cavernous transformation (the “pseudocholangiocarcinoma sign”) should be considered an adaptive change with a low degree of clinical importance. A wide spectrum of clinical findings can be seen, ranging from normal physical and biochemical findings, to overt cholestasis with liver injury.^{10 12 13} The term ‘portal ductopathy’ has been used to reflect changes in biliary ducts in a better manner, which in the long term, result in biliary stasis, jaundice, cholangitis and cholelithiasis, adding to morbidity and, possibly, mortality.^{10 11}

Selection of appropriate patients for intervention remains an important issue. Ramos *et al*² reported a case of a 58-year-old man with cavernous transformation of the portal vein secondary to PVT who presented with anaemia, electrolyte imbalance and deranged LFTs. In this case, no intervention was offered as the patient was considered a poor candidate for an invasive procedure and the LFTs improved spontaneously. This raises the possibility of spontaneous biochemical improvement in patients without visible filling defects or strictures.

Aguirre *et al*¹⁴ reported a case series of 11 patients with cavernous transformation of the portal vein who were followed up with clinical imaging. Of these 11 patients, all had elevated LFTs on initial presentation. On follow-up, there was no additional rise in LFT levels, and no progression in biliary tree dilatation. Significantly, no patient required endoscopic or surgical intervention, and no correlation was found between degree of bile duct dilatation, severity of portal cavernous transformation and LFT levels.

Several case studies describe the use of ERCP and stents in biliary tract pathology on the background of cavernous transformation of the portal vein.^{8 15–17} Although endoscopic treatment has generally been selected as the first treatment option, some cases still required surgical intervention.^{8 16 17}

One case study reported by Perego *et al*⁸ involved treatment of choledocholithiasis secondary to a stricture of the CBD in a patient with cavernous transformation of the portal vein. Stone removal via ERCP was attempted several times, but a percutaneous transhepatic approach was eventually required with stone removal and pneumatic dilation of the stricture. Bejanin *et al*¹⁵ reported a case series of 10 patients, who all had severe biliary strictures, treated with biliary sphincterotomy and stent insertion. Four patients displayed improvement in their biliary stricture. Mork *et al*¹⁶ presented a report of two cases of cavernous transformation of the portal vein associated with a CBD stricture. Both cases were initially treated with ERCP, which was successful in one case; the other was eventually referred for surgical management.

On review of the available literature, endoscopic management appears to be a safe first-line option in the treatment of symptomatic portal biliary ductopathy associated with cavernous transformation of the portal vein. Surgical intervention may be necessary in some cases in which sphincterotomy, balloon dilatation and/or stent insertion are unsuccessful.^{8 16}

Learning points

- ▶ Symptomatic biliary obstruction on a background of portal biliary ductopathy caused by cavernous transformation of the portal vein may occur in the absence of strictures or filling defects.
- ▶ Portal biliary ductopathy in the setting of cavernous transformation of the portal vein can be safely and effectively treated endoscopically.
- ▶ The spectrum of disease is broad, and patients may demonstrate improvement, regardless of intervention.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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