CASE REPORT

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Therapeutic Challenges for Symptomatic Portal Cavernoma Cholangiopathy

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ABSTRACT

Although transjugular intrahepatic portosystemic shunts are most frequently used for the management of portal hypertension, the surgical approach is preferred for symptomatic portal cavernoma cholangiopathy. We present the case of a 25-year old female patient with a portal cavernoma secondary to catheterization of the umbilical vein at birth. She had had two episodes of esophageal variceal bleeding, successfully treated by endoscopic banding. and an episode of acute cholangitis secondary to portal cavernoma cholangiopathy. Endoscopic sphincterotomy and biliary stenting were performed, and were followed by repeated episodes of biliary stent occlusion. The last biliary drainage procedure triggered a massive hemobilia. Since endoscopic therapy was ineffective, a surgical mesocaval shunt with graft interposition and splenectomy was performed with favorable outcome. In selected cases, the mesocaval shunting plays an essential role in the treatment of portal cavernoma cholangiopathy even in the era of interventional radiology.

Key words: portal cavernoma – portal cavernoma cholangiopathy – portosystemic shunts – mesocaval surgical shunt.

Abbreviations: CBD: common bile duct; CT: computed tomography; ERCP: endoscopic retrograde cholangiopancreatography; IVC: inferior vena cava; LHD: left hepatic duct; MRCP: magnetic resonance cholangiopancreatography; PC: portal cavernoma; PCC: portal cavernoma cholangiopathy; SMV: superior mesenteric vein.

INTRODUCTION

Portal cavernoma (PC) represents a porto-portal collateral pathway that serves as a bypass through the thrombosed vein. Its major consequences are represented by portal hypertension and anomalies within the biliary tree [1-3]. Portal cavernoma cholangiopathy (PCC) is defined as abnormalities in the extrahepatic biliary system including the cystic duct and gallbladder, with or without abnormalities in the first and second generation biliary ducts, in a patient with portal cavernoma in the absence of other causes [4, 5]. Previously used names were portal hypertensive biliopathy, portal biliopathy or portal cholangiopathy [6]. The biliary obstruction has two components: a reversible one, i.e. compression by engorged collaterals, and an irreversible one, due to the fibrotic stenosis of bile duct wall induced by ischemia and inflammation [7].

Portal cavernoma is a mandatory prerequisite in the diagnosis of PCC. Magnetic resonance cholangiopancreatography (MRCP) combined with an MR portovenography depicts biliary and vascular abnormalities. Endoscopic retrograde cholangiopancreatography (ERCP) is an invasive procedure, reserved only for therapeutic purposes such as biliary stenting or extraction of bile duct stones [7, 8].

Decompression of the bile ducts and the portal vein system are the main objectives of PCC treatment [9]. Optimal management is reached by the close coordination between skilled endoscopic and surgical teams with sequential interventions designed to establish and maintain biliary drainage, then to decompress the portal cavernoma, and finally, if required, second stage biliary surgery or endotherapy for biliary strictures [10].

CASE REPORT

Our patient, a 25-year old female, experienced a cerebral hemorrhage at birth complicated by chronic encephalopathy with mental retardation, spastic tetraparesis and epilepsy. At the

age of 18 she had two episodes of esophageal variceal bleeding, which were treated by endoscopic variceal banding. The Doppler ultrasound examination of the portal vein revealed a portal cavernoma, secondary to the catheterization of the umbilical vein at birth.

At 24 years of age the patient was admitted to the Emergency Department with biliary colic, obstructive jaundice and fever. Laboratory tests revealed mild cholestasis, hyperbilirubinemia and elevated aminotransferases. The abdominal Doppler ultrasound confirmed the presence of the PC located both at the hilum and in the liver, along with the dilation of the intrahepatic bile ducts in both lobes, splenomegaly, and collateral circulation in the spleen hilum and the gallbladder wall (Fig. 1). A clinical diagnosis of acute cholangitis secondary to common bile duct (CBD) obstruction was established. The ERCP aspect was pathognomonic for PCC with a dominant stenosis in the middle portion of the CBD (Fig. 2), stented with a 10 Fr/9 cm plastic prosthesis. In the following six month period, the patient experienced five biliary stent occlusion episodes complicated by cholangitis. The biliary endoscopic drainage procedures were regularly complicated by hemobilia. An oval filling defect in the cholangiographic image was considered a stone instead of a pericholedochal varix and the maneuvers with the Dormia basket caused hemobilia (Fig. 3). During the last ERCP procedure the hemobilia was severe (Fig. 4) with marked anemia, requiring blood transfusion and terlipressin therapy. A CT angiography confirmed the PC and the patency of the superior mesenteric vein (SMV), splenic vein and inferior caval vein (ICV) (Fig. 5).

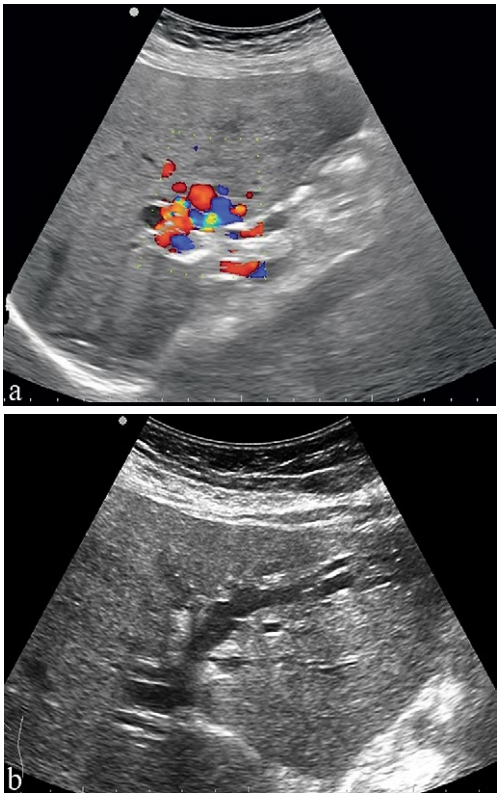

Fig. 1 Ultrasonography before surgery: multiple tortuous periportal veins in the hepatic hilum corresponding to the portal cavernoma (1a) and dilatation of the intrahepatic bile duct radicles (1b).

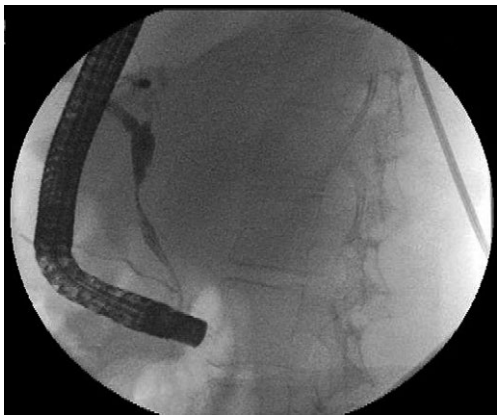

Fig. 2. Endoscopic retrograde cholangiography images: stricture in the middle portion of the common bile duct and upstream dilatation, irregular contour and extrinsic impression on the bile ducts.

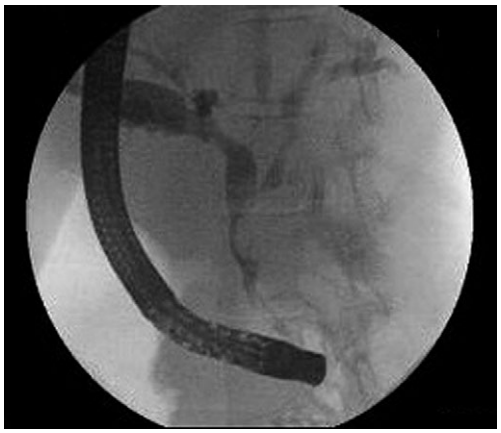

Fig. 3. ERCP appearance misleading the varix with a stone (oval shaped filling defect in the common bile duct).

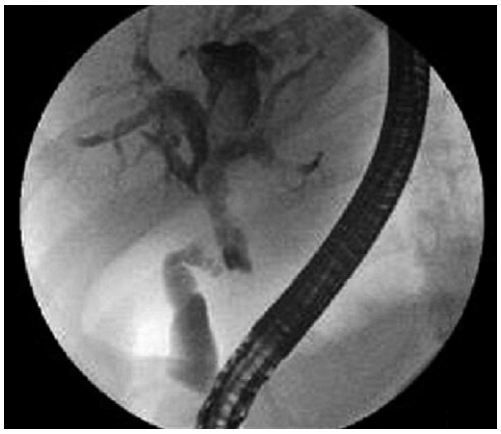

Fig. 4. Endoscopic retrograde cholangiography aspect of hemobilia with multiple filling defects in the bile ducts (clots).

A surgical side-to-side mesocaval shunt with Goretex graft interposition and splenectomy were performed. The postoperative evolution was favorable with the resolution of the cholangitis and the episodes of hemobilia, normalization of bilirubin and transaminase levels, and reduction of cholestasis

enzymes levels. The Doppler ultrasound performed at the postoperative check-up at one year showed regression of the cavernoma, no intrahepatic bile duct dilatation and optimum function of the mesocaval shunt (Vmax = 80 cm/sec) (Fig. 6). Currently, the patient is under anticoagulant therapy with vitamin K antagonists, anti-platelet drugs (aspirin 75 mg/ day), ursodeoxycholic acid 10 mg/ kg/day, and anticonvulsants (phenobarbital 100 mg/day).

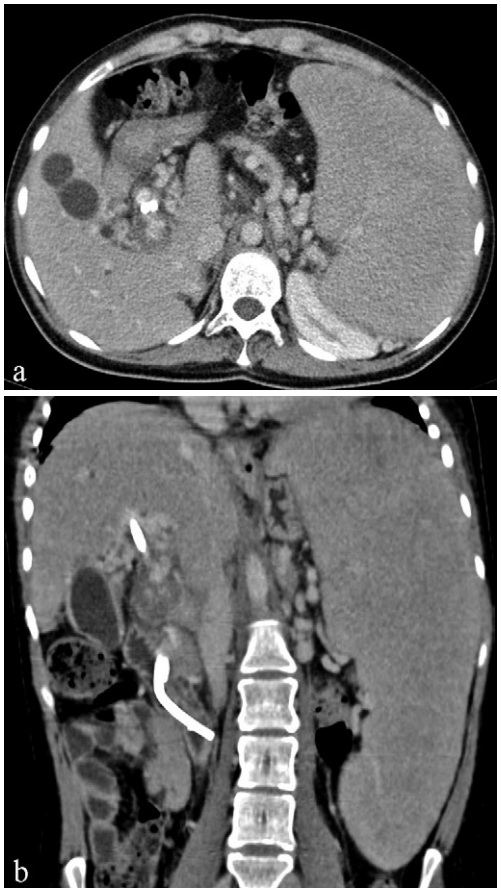

Fig. 5. CT angiography with axial view (4a) and coronal view (4b) shows the portal cavernoma in the hepatic hilum, the two biliary stents, the splenomegaly and the portosystemic collaterals.

DISCUSSION

Portal cavernoma cholangiopathy occurs in 81-100% of the patients with PC, but it is symptomatic in only 5-38% of cases. It has a slow progression and a natural history divided into four stages: preclinical, asymptomatic, symptomatic and complicated [11]. Portal cavernoma represents the preclinical stage of PCC and it is often asymptomatic. A careful evaluation will, however, reveal elements of portal hypertension, such as gastroesophageal varices, collateral circulation and splenomegaly. Variceal bleeding represents the first clinical manifestation of PC in 20-40% of cases [12, 13]. In the asymptomatic stage, which has an average duration of 8-10 years, early changes can already be identified at MRCP (irregularities, parietal impressions, stenosis) with or without altered biochemical tests (minor increase in alkaline phosphatase and bilirubin levels) [14]. Biliary colic, jaundice, cholestasis and cholangitis appear late in the evolution of the disease and only in patients with advanced changes at MRCP [8]. Risk factors for the occurrence of symptomatic PCC are: prolonged duration of the disease, history of variceal hemorrhage, cholestasis, bile duct stones and advanced (grade III) cholangiographic findings (biliary strictures, dilatations) [15]. The complicated stage includes patients with advanced bile duct changes and progressive liver dysfunction (secondary biliary cirrhosis), and patients with limited therapeutic options [11].

Our patient followed the sequence of clinical events similarly described in the literature: catheterization of umbilical vein at birth with thrombosis of the portal vein, later transformed into a cavernoma, variceal hemorrhage at 18 years of age as the first clinical manifestation of PC and repetitive episodes of acute cholangitis at the age of 24 representing the symptomatic phase of PCC.

The Doppler ultrasound and CT angiography confirmed the PC (periportal spongiform network of venous-type vessels) along with its consequences: portal hypertension (splenomegaly, collateral circulation in the hilum of the spleen and gallbladder wall), associated with cholangiopathy (intrahepatic bile duct dilatation). Although MRCP associated with MR portovenography represents the standard for PCC

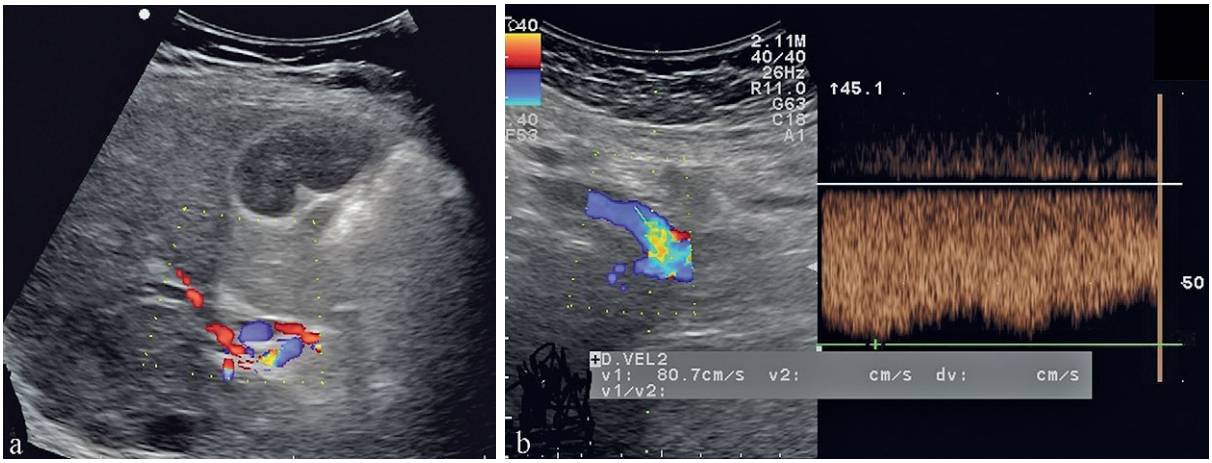

Fig. 6. Postoperative Doppler ultrasonography reveals the regression of the cavernoma (6a), the patency of the mesocaval shunt and the maximum velocity inside the shunt (6b).

diagnosis [16], in this particular situation, ERCP was preferred, serving both as a diagnostic and a therapeutic procedure. The ERCP confirmed grade III biliary changes [17]. The filling defect given by an intracoledochal varix was mistaken for a coledochal stone and the maneuvers of the Dormia basket, meant to remove the suspected stone, triggered hemobilia.

The treatment of symptomatic PCC should be focused on the therapy of cholangitis (biliary drainage + antibiotic therapy), followed by the decompression of the portal venous system (portosystemic shunts) [18]. Endoscopic stenting is the first line of intervention in cases of cholangitis secondary to biliary strictures. Hemobilia and cholangitis, which occur in 30% of bile stent replacement procedures, are the major complications of endoscopic therapy [10]. Decompression of the portal venous system has several advantages: it reduces the PC size, alleviates the biliary obstruction, reduces the size of the esophageal varices, decreases the risk of variceal bleeding, and corrects the hypersplenism [10]. End-to-side portacaval shunts pose the risk of liver failure and of encephalopathy and have been abandoned in favor of alternatives that maintain a proper hepatic perfusion: mesocaval shunts with graft interposition, distal splenorenal Warren shunts and side-to-side portocaval calibrated shunts [19]. The first use of a mesocaval surgical shunt dates back to 1963 and has since become the first choice treatment of PC complicated by variceal hemorrhage or by PCC. The interposition of a graft between the superior mesenteric vein and the inferior vena cava improved the performance of the shunt [20, 21]. A correct graft diameter (<14 mm) is of particular importance as it ensures proper hepatic perfusion as well as the prevention of hepatic encephalopathy [22, 23]. Traditional surgical portosystemic shunts are associated with a high operative morbidity, which has spurred the development of minimally invasive radiology techniques - transjugular intrahepatic portosystemic shunt, direct intrahepatic portacaval shunt and percutaneous mesocaval shunt [24]. Patients with PC would not routinely be candidates for TIPS. Percutaneous mesocaval shunt creation may be considered in cases of PC with a patent mesenteric vein [24].

In our patient, endoscopic biliary drainage was ineffective and was followed by hemobilia and cholangitis. Decompression of the portal venous system was envisaged. TIPS was not indicated due to the presence of PC. Although a percutaneous mesocaval shunt would have been the best minimally invasive decompression option, this procedure was not available. There were also some theoretical drawbacks, which precluded its use: narrow clinical experience, intraperitoneal hemorrhage secondary to the vessels puncture and inadvertent bowel perforation. The surgical procedures carried out were mesocaval shunt with Goretex graft interposition and splenectomy.

The favorable postoperative outcome with reduction in the PC size and the resolution of the biliary obstruction demonstrates the beneficial role of mesocaval shunting in the therapy of PCC.

CONCLUSION

Endoscopic biliary drainage and surgical decompression of the portal venous system are complementary in the management of patients with symptomatic PCC. Nonetheless, although interventional radiology is minimally invasive, mesocaval surgical shunting continues to play an important role in the therapy of this disease.

Conflicts of interest: None to declare.

Authors' contributions: A.C.: ultrasonography monitoring, clinical management and manuscript writing; I.P.: surgical intervention of the patient, literature search and manuscript reviewing; V.M.: ERCP investigation, manuscript reviewing; O.A.: CT angiography. All authors approved the final version of the manuscript.

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Posibilități terapeutice ale colangiopatiei simptomatice associate cu cavernomul portal

ABSTRACT / REZUMAT

Deși șunturile portosistemice transjugulare sunt frecvent utilizate la pacienții cu hipertensiune portală, abordarea chirurgicală este preferată in cazurile de colangiopatie asociată cavernomului portal simptomatic.

Prezentăm cazul unei paciente de 25 de ani cu cavernom portal secundar cateterizării venei ombilicale la naștere. A prezentat două episoade de sângerare variceală, tratate eficient endoscopic prin montare de ligaturi elastic, și o angiocolită acuta în contextul colangiopatiei asociate cavernomului portal simptomatic. Sfincterotomia endoscopică asociată cu stentarea căii biliare principale a constituit abordarea inițială, dar a fost complicată cu repetate episoade de colmatare a protezei. Ultima procedură de drenaj biliar a fost insoțită de hemobilie masivă. Având in vedere ineficiența terapiei endoscopice s-a optat pentru o abordare chirurgicală reprezentată de șunt mezenterico-cav cu interpoziție de grefă vasculară și splenectomie. Postoperator, evoluția a fost favorabilă.

In cazuri selectate, șunturile mezenterico-cave joacă un rol esențial în tratamentul pacienților cu colangiopatie asociată cavernomului portal.