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CASE REPORT | ENDOSCOPY

Successful Hemostasis Using Fully Covered Self-Expanding Metallic Stent for Spontaneous Hemobilia in a Child With Portal Cavernoma Cholangiopathy

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ABSTRACT

Portal cavernoma cholangiopathy refers to changes in the intrahepatic and extrahepatic biliary ducts in patients with extrahepatic portal venous obstruction. Spontaneous hemobilia in the setting of portal cavernoma cholangiopathy is extremely rare, and it poses diagnostic as well as therapeutic challenge. Here, we report the case of a 10-year-old girl with extrahepatic portal venous obstruction, who presented with hemobilia. Computed tomography angiography of abdomen and endoscopic ultrasound confirmed the presence of pericholedochal, paracholedochal, and intracholedochal varices. Hemostasis was achieved with the placement of a fully covered self-expanding metallic stent into the common bile duct. Fully covered self-expanding metallic stent is safe and effective for control of bleeding in children presenting with hemobilia.

KEYWORDS: spontaneous hemobilia; portal cavernoma cholangiopathy; fully covered self-expanding metallic stent

INTRODUCTION

Portal cavernoma cholangiopathy (PCC) is one of the most dreaded complications of extrahepatic portal venous obstruction (EHPVO). PCC refers to the abnormalities of extrahepatic and intrahepatic bile ducts due to compression by collaterals.[1] Although PCC in children with EHPVO is as common as that in adults, symptomatic PCC is uncommon (7%).[1,2] Spontaneous hemobilia due to rupture of choledochal varices into the biliary system has been reported in adults.[3,4] Pediatric literature is lacking. Here, we report a case of a child with EHPVO who presented with spontaneous hemobilia and was successfully managed with a fully covered self-expanding metallic stent (FCSEMS).

CASE REPORT

A 10-year-old girl with EHPVO had variceal bleeding at the age of 6 years. She underwent endoscopic variceal ligation for grade III esophageal varices (Paquet grading) at first endoscopy, and downgrading of esophageal varices was achieved over next 8 months.[5] She was lost to follow-up for the next 3 years, and then was readmitted with hematemesis due to recurrence of varices and underwent endoscopic sclerotherapy (EST) for grade II esophageal varices. Three weeks after EST, she presented again with 2 days of hematemesis and melena. There was no history of pain abdomen, fever, nonsteroidal anti-inflammatory drug intake, jaundice, or abdominal trauma. On admission, the patient was pale and had tachycardia (heart rate 146/bpm, blood pressure 96/60 mm Hg). She had anemia (hemoglobin 5.6 g/dL), thrombocytopenia (78,000/mm^3), and normal total leukocyte count (4,300/mm^3). Liver function tests were as follows—total/direct bilirubin: 2.4/0.9 mg/dL, aspartate/alanine transaminase: 32/38 U/L (normal: 5–40 U/L), alkaline

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phosphatase: 586 U/L (normal 35–150 U/L), gamma-glutamyl transpeptidase: 180 U/L (normal 13–86 U/L), albumin: 3 g/dL, and international normalized ratio: 1.2. She was started on intravenous fluids and octreotide infusion, and packed red blood cell transfusion was given. Upper gastrointestinal endoscopy at ~6 hours from admission showed grade I esophageal varices with a post-EST ulcer, no gastric varices, normal duodenum, and no ongoing bleed. Proton pump inhibitor infusion was started. However, the child had a repeat bout of hematemesis after 36 hours with a significant drop in hemoglobin. Repeat endoscopy showed blood coming out of the major duodenal papilla, suggestive of hemobilia (Figure 1). Computed tomography angiography of abdomen was done, which revealed extensive pericholedochal collaterals, causing extrinsic compression of the common bile duct (CBD) and a shuntable anatomy (patent splenic vein [diameter 7 mm], superior mesenteric vein [6.4 mm], left renal vein [5.8 mm]). Endoscopic ultrasound (EUS; Olympus GF-UCT180, EVIS EXERA II ultrasound gastro-videoscope; Olympus Inc, Tokyo, Japan) confirmed the presence of multiple, large pericholedochal, paracholedochal, and intracholedochal collaterals (no perforator); however, these were not amenable for glue injection due to the large number of vessels (Figure 2). To stop ongoing bleeding, endoscopic retrograde pancreato-cholangiography was performed using standard adult duodenoscope (Olympus TJF 160R; Olympus), which showed active bleeding from the major papilla. The cholangiogram showed indentation near lower and mid-CBD, suggestive of intracholedochal varices and a CBD diameter of 5 mm. A 10 mm × 60 mm FCSEMS (Wallflex Biliary RX fully covered stent system; Boston Scientific, MA) was placed into the CBD (Figure 3) after a small sphincterotomy (2–3 mm) to facilitate passage of stent. Another double pigtail plastic stent (7 Fr × 5 cm) was placed inside the FCSEMS to prevent its migration (Figure 3). Hemobilia stopped, with no further drop in hemoglobin and no complications. The patient was discharged after 7 days. The child underwent

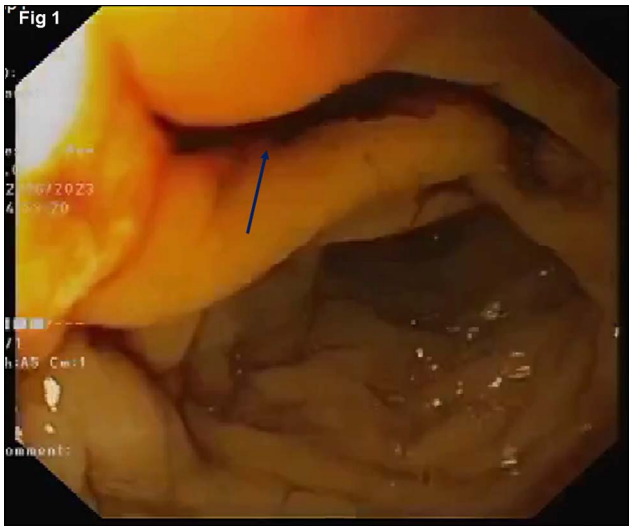

Figure 1. Upper gastrointestinal endoscopy image showing blood coming out of papilla (blue arrow), indicating hemobilia.

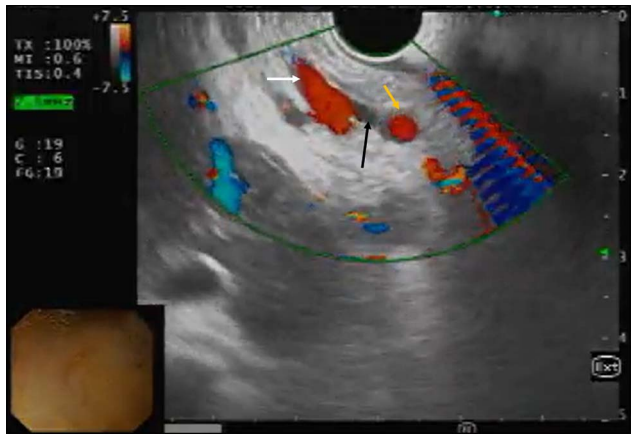

Figure 2. Endoscopic ultrasound images showing intracholedochal (yellow arrow) and pericholedochal varices (white arrow) around the common bile duct (black arrow).

splenectomy with proximal splenorenal shunt 3–4 weeks after the bleed. Unfortunately, the shunt was blocked at 3 months after surgery, and she is awaiting radiological intervention for reopening of the shunt at the last follow-up. The patient did not have any recurrence of bleed, and the metallic stent is still in situ.

DISCUSSION

Hemobilia in the setting of PCC has been reported after instrumentation such as percutaneous transhepatic biliary drainage, transjugular intrahepatic portosystemic shunt, post-liver biopsy or sphincterotomy, balloon sweeping, and stenting during endoscopic retrograde pancreato-cholangiography.[6–11] Spontaneous hemobilia from PCC is an extremely rare phenomenon. Till now, only 2 cases of spontaneous hemobilia from PCC have been reported in adults.[3,4] To the best of our knowledge, this is the first report of spontaneous hemobilia in a child with PCC.

The risk of hemobilia in PCC is related to the presence of pericholedochal and intracholedochal collaterals and congestion of vessels in the CBD wall. In particular, the transient pressure elevation in the biliary varices during balloon sweeping can increase the risk of bleeding.[1,6,10]

The diagnosis of hemobilia as the source of upper gastrointestinal bleeding can be easily overlooked because it is intermittent in nature. The classical triad of right upper quadrant pain, jaundice, and gastrointestinal bleed is found in only one-third of patients.[12] In children, a history of pain abdomen may not always be present, similar to our case. Clinical suspicion should hinge on the presenting symptoms and clues in medical history even if all the typical symptoms are not present and no history of instrumentation is found.

Doppler ultrasound may reveal mobile or hypoechoic, non-shadowing filling defects in gall bladder or CBD. Computed

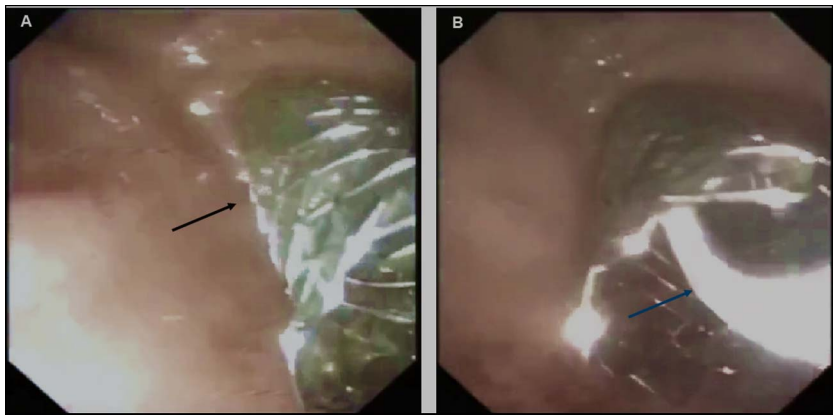

Figure 3. Side-viewing endoscopic images showing successful placement of a 10 mm × 60 mm fully covered self-expanding metallic stent (black arrow) into common bile duct, resulting in bleeding control (A). A double pigtail plastic stent (7 Fr × 5 cm) (blue arrow) was placed inside the metallic stent to prevent its migration (B).

tomography angiography is helpful as it evaluates the vascular anatomy in addition to the hepatobiliary system and helps in planning endovascular interventions.[11] EUS is a useful tool for both diagnostic evaluation and treatment in cases of PCC.[4] In our case, EUS confirmed the diagnosis of PCC and helped in deciding further treatment plan.

Minor hemobilia may be managed with conservative therapy, whereas massive hemobilia can be potentially life-threatening and require invasive treatment. While existing methods include balloon tamponade and endovascular embolization techniques, endoscopic hemostasis can be achieved by tamponade effect through FCSEMS deployment.[13] FCSEMS deployment in CBD provides local tamponade and secures hemostasis, as was seen in our case where medical therapy failed to control the bleeding. It was a life-saving measure for our child. After initial hemostatic measures, definitive treatment for underlying etiology such as shunt surgery in EHPVO should be performed to prevent recurrence of hemobilia.

Stent migration is the most common complication of FCSEMS, seen in 20%–40% of cases.[14–16] To prevent this, an additional plastic stent is placed within the FCSEMS, similar to our case.[17,18] Cholangitis and/or cholecystitis can occur due to obstruction of one of the bile ducts by the proximal covered end of the FCSEMS. Hence, use of smaller length FCSEMS has been advocated to prevent these complications, as was done in our case.[19]

Our case highlights the importance of recognizing hemobilia as a rare cause of upper gastrointestinal bleeding in children with EHPVO and PCC and utility of FCSEMS in securing hemostasis.

Hemobilia is an important complication of PCC. FCSEMS is feasible, effective, and safe for securing hemostasis in children with hemobilia.

DISCLOSURES

Author contributions: A. Samanta did the literature review, A. Samanta and A. Srivastava co-drafted the initial manuscript. A. Samanta, A. Srivastava, U. Poddar, MS Sarma, B. Kumar were involved in the clinical management of the case, S. Mohindra did endoscopic ultrasound and endoscopic retrograde pancreato-cholangiography, R. Yadav interpreted the radiological images, B. Kumar did the surgical procedure. U. Poddar, MS Sarma, S. Mohindra, BK and R. Yadav have critically reviewed the manuscript and provided inputs. All authors approved the final version of the submitted manuscript. A. Srivastava is the guarantor of this article.

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