Massive gastrointestinal bleeding due to isolated jejunal varices in a patient without portal hypertension

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ABSTRACT

INTRODUCTION: Isolated ectopic varices located in the small bowel are uncommon. Portal hypertension caused by liver cirrhosis is the most common predisposing risk factor.

PRESENTATION OF CASE: We present an unusual case of massive gastrointestinal bleeding from idiopathic jejunal varices in a 73-year-old Caucasian male without portal hypertension. Exploratory laparotomy disclosed ectopic varices located in the small intestine. Segmental resection of the jejunum with end to end anastomosis resulted in a complete resolution of the haemorrhage. During a 5 year follow up, the patient is stable with no bleeding recurrence.

DISCUSSION: Information on aetiology, diagnosis and management of jejunal varices is reviewed.

CONCLUSION: Diagnosis and management of isolated jejunal varices is challenging. Surgeons as well as acute care physicians have to consider idiopathic form of jejunal varices as a potential cause of gastrointestinal bleeding when gastroduodenoscopy and colonoscopy are negative.

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1. Introduction

Ectopic varices located in the small intestine are uncommon and are usually caused from portal hypertension secondary to hepatic cirrhosis. 1 Jejunal variceal bleeding has been rarely reported 1 and represents a serious diagnostic dilemma. 2 Here we present an atypical case of a 73 year old man with massive gastrointestinal bleeding due to isolated jejunal varices in the absence of portal hypertension.

1.1. Case presentation

A 73 year old Caucasian male was referred by his general practitioner to the emergency department of the Saint George General hospital of Chania, Crete due to one episode of hematochezia, hematemesis and abdominal pain. His past medical history was unremarkable except for chronic atrial fibrillation treated with acenocoumarol and cholecystectomy for gallstone 10 years previously. His vital signs on admission were as follows: blood pressure, 80/40 mmHg; heart rate, 120 beats/min; oxygen saturation, 98% while he was breathing ambient air.

Initial laboratory work up disclosed: white blood count, 5.20 K/µL (range: 4–11 K/µL); hematocrit (Hct), 27.1% (range: 40–50%); haemoglobin (Hb) 9.2 g/dl (range: 13.5–17.5 g/dl); platelet counts (PLT), 119 K/µl (range: 150–450 K/µl); urea 76 mg/dl (range: 10–50 mg/dl); creatinine 0.7 mg/dl (0.7–1.2 mg/dl); glucose, 153 mg/dl (60–115 mg/dl); sodium 146 mmol/L (range: 137–147 mmol/L); potassium, 4.8 mmol/L (3.5–5.5 mmol/L); aspartate aminotransferase (AST), 13 U/L (range: 0–38 U/L); alanine-aminotransferase (ALT), 22 U/L (4–36 U/L); total protein, 3.7 g/dl (6.4–8.3 g/dl); serum albumin, 3.4 g/dl (range: 6.4–8.3 g/dl); total bilirubin 4.8 g/dl (range: 0–1.1 g/dl). Blood coagulation tests showed: prothrombin time (PT), 20 sec (range: 9.8–15 sec); activated partial thromboplastin time (aPTT), 31 sec (26–36s); International Normalized Ratio (INR), 1.69 (range: 0.85–1.15). Serological markers for previous or current hepatitis B and C infection were negative. There was no previous history of alcohol abuse. Digital rectal examination disclosed red bright stools on the glove. Abdominal examination showed a marked sensitivity on palpation of the epigastric area. During hospital stay the patient presented haemodynamic instability (Hct, 19.1%; Hb, 6.5 g/dl; PLT, 88 K/µL) and required transfusion with totally five units of red blood cells.

Endoscopic evaluation with colonoscopy and esophagogastroduodenoscopy was negative for active source of bleeding. Abdominal aortic angiography was not feasible due to an episode of hematemesis during the procedure along with a simultaneous drop of patient’s blood pressure. Urgent exploratory laparotomy was consecutively performed and revealed dilated veins extended on a 12 cm surface of the proximal jejunum, 15 cm beyond the ligament of Treitz (Fig. 1). There were no findings of arterio-venous fistula or other malformation. Liver was normal sized without
evidence of cirrhosis. Ascitic fluid was not present. After identification of the bleeding site (Fig. 2) resection of the involved jejunal segment with end to end anastomosis was performed (Fig. 3). Histological examination of the resected specimen showed enlarged calibre veins located in the submucosa. Liver biopsy was negative for cirrhosis. The post-operative course was uneventful and the patient was discharged home one week after surgery. At 5 years follow up the patient was stable with no further complications.

2. Discussion

Bleeding due to isolated jejunal varices represents an extremely rare cause of gastrointestinal haemorrhage with potential fatal outcomes if not correctly diagnosed and treated. Remarkably, eleven cases have been described until 1992, clinically manifested with melena or hematochezia. Aetiology of jejunal variceal formation is not completely clarified. Common predisposing risk factors are hepatic cirrhosis with portal hypertension, hepatocellular carcinoma, pancreatitis. A familial aggregation of small intestinal varices in the absence of portal hypertension has been described. Intra-abdominal adhesions from previous abdominal surgery, arteriovenous fistula secondary to trauma and nodular lymphoid hyperplasia have been also reported.

Jejunal varices cause significant difficulties in diagnosis and management. The classical clinical triad that characterizes symptomatic ectopic jejunal varices consists of portal hypertension, hematochezia without hematemesis and past abdominal operation. Capsule endoscopy has been reported to be a useful non-invasive diagnostic tool for patients with obscure gastrointestinal bleeding including those with chronic hepatic diseases. Surgical management includes segmental jejunal resection with anastomosis, transjugular intrahepatic portosystemic shunts (TIPS) and variceal ligation. Insertion of TIPS and balloon-occluded retrograde transvenous obliteration represent interventional radiologic modalities indicated in patients with a poor condition. Percutaneous paraumbilical embolization via caput medusae could be useful in cirrhotic patients with contraindications to surgery.

In the case presented, jejunal varices were formatted in the absence of portal hypertension. Although previous abdominal surgery was the only predisposing risk factor, no arteriovenous fistula or malformation was visualized during exploratory laparotomy. Idiopathic ectopic varices located in the jejunum represent a very uncommon clinical occurrence. Furthermore, hematemesis as an initial clinical manifestation of jejunal variceal bleeding is uncommon. Accurate diagnosis of jejunal varices represents a challenging issue. Since fatal clinical conditions rarely allow diagnostic revisions we conclude that acute care physicians have to consider the idiopathic form of jejunal varices in the differential diagnosis of gastrointestinal haemorrhage with negative findings from upper and lower endoscopy.

Conflict of interest declaration

The authors declare that they have no competing interests.

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Informed consent

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Author contributions

Nikolaos Katsougris and Emmanouil Bobolakis performed the operation and provided substantial information regarding the patient’s case. Dimitrios Anyfantakis was involved with the primary care of the patient. Dimitrios Anyfantakis and Miltiades Kastanakis had a major contribution in the preparation and revision of the manuscript. All authors have read and approved the final manuscript.

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