CASE REPORT

Ectopic varices masquerading as lower gastrointestinal bleeding

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SUMMARY

A middle-aged woman previously in good health arrived to hospital with severe haemorrhagic shock. An abdominal and pelvic CT scan showed hepatomegaly, fluid in the sigmoid colon, perigastric and right sided pelvic varices without a clear source of bleeding. Urgent esophagogastroduodenoscopy excluded gastric varices and active upper gastrointestinal bleeding and the colonoscopy was inconclusive due to haemodynamic instability and massive bleeding which made it impossible to proceed with a full colonoscopy. An explorative laparotomy identified liver cirrhosis and bleeding from varices of the right fallopian tube which had formed a fistula with the caecum. A right hemicolectomy with a side to side anastomosis and a right sided salpingo-oophorectomy were carried out. The patient was thereafter transferred to the intensive care unit intubated, requiring massive fluid resuscitation and high doses of vasoactive agents. Nine days after hospital admission she was transferred to the surgical ward and followed up by a hepatologist.

of 48 g of alcohol a day was reported. On arrival she was pale with cold extremities, with a resting tachycardia (120 bpm) and a systemic blood pressure of 110/70 mmHg. Initially the Glasgow Coma Score was 15/15. The haemoglobin level on arrival was 102 g/L with a mean cell volume of 100 fl, platelet count of $156 \times 10^9/L$, the coagulation profile was within normal range and arterial serum lactate was 5.6 mmol/L. The liver function illustrated elevated alkaline phosphatase of 139 U/L, Gamma-glutamyl-transferase of 531 U/L and aspartate transaminase of 77 U/L. She had a distended abdomen with mild tenderness on palpation of the lower quadrants, bowel sounds were present and profuse fresh rectal bleeding with blood clots was noted. On physical examination she had multiple large spider naevi mostly on the lower limbs. The patient started to quickly deteriorate in the emergency department with a worsening neurological status, rigors and fever. Urgent blood transfusion and fresh frozen plasma were administered and she was taken for an urgent contrast enhanced CT abdomen and pelvis to identify the source of bleeding.

BACKGROUND

Portal hypertension leading to the formation of gastrointestinal varices is an established complication frequently seen in liver cirrhosis and screening in asymptomatic patients is a recognised practice. Furthermore the literature reports that 5% of variceal bleeding results from ectopic varices.^{1–9} Common sites of ectopic variceal formation are: duodenum, jejunum, ileum, colon, rectum, biliary tree, ovary and vagina.⁷ This article describes a patient with an unusual first presentation of her cirrhosis complicated by fistula formation between right fallopian tube varices and caecum demonstrating that varices in the pelvic venous circulation is a potential complication which could be life threatening.

CASE PRESENTATION

A middle-aged woman presented to the emergency department of the Regional Hospital of Mendrisio, Switzerland, with stage 2 haemorrhagic shock caused by a suspected lower gastrointestinal bleeding. The patient denied any significant medical history. She reported self-administering over-the-counter acetylsalicylic acid and a beta blocker for suspected hypertension. On arrival she had several episodes of fresh rectal bleeding associated with dizziness. She denied any abdominal pain, vomiting, fever or syncope and regular alcohol consumption

INVESTIGATIONS

The abdominal CT scan illustrated hepatomegaly with an inhomogeneous aspect, perigastric and right sided pelvic varices (figure 1) with fluid present in the sigmoid colon and the rectum. Angiography was not available and the contrast enhanced CT scan was not able to identify bleeding from a specific site during the arterial and venous phases. The literature indicates that even when multidetector CT angiography is carried out the exact source of bleeding may not always be identified as the bleeding can be intermittent from minute to minute.¹⁰ Considering that the CT scan was inconclusive it was necessary to proceed with invasive measures. The patient was taken first to intensive care unit (ICU) for haemodynamic stabilisation requiring further blood products (1 unit of packed red blood cells, 2 units of fresh frozen plasma, 2 g of fibrinogen, 1 unit of platelets and 1000 units of prothrombin-complex concentrates), fluid resuscitation with crystalloids and support with norepinephrine. During the haemodynamic stabilisation phase, she developed high fever (39°C) and blood and urine cultures were carried out. Shortly afterwards she was taken to the operating theatre where an esophagogastroduodenoscopy excluded gastro-oesophageal varices, gastric varices and upper gastrointestinal bleeding. Afterwards a colonoscopy was attempted, as angiography was not available, and it was only possible

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Figure 1 Abdominal CT scan: frontal view contrast enhanced. CT scan demonstrating suspected ectopic varices in the perigastric region (upper arrow) and the right pelvic region (lower arrow).

to visualise the colon up to the hepatic flexure. Due to haemodynamic instability and ongoing massive blood loss it was not possible to proceed with a full colonoscopy and endoscopically guided therapeutic measures. Considering the difficulty in identifying the source of bleeding the patient underwent an explorative laparotomy.

DIFFERENTIAL DIAGNOSIS

After excluding upper gastrointestinal bleeding, the differential diagnoses included sources of bleeding from the lower gastrointestinal tract: diverticulosis and diverticulitis, angiodysplasia, inflammatory bowel disease and ectopic varices.

TREATMENT

The explorative laparotomy identified an enlarged cirrhotic liver with moderate ascites and there were no gastric varices identified on outside inspection of the stomach.

In the pelvic region there was a collateral venous system originating from the right fallopian tube which passed through the omentum and an epiploic appendix connecting to the venous system of the caecum. A short distance from the ileocecal valve bleeding into the colon was identified in keeping with the fistula formation (figure 2). A right hemicolectomy with a side to side anastomosis, right sided salpingo-oophorectomy and partial omentectomy were carried out. Inside the excised ascending colon approximately 500 mL of blood were estimated. The histopathological report identified numerous and large vascular ectasia in the submucosa of the caecum, at the level of the right ovary, right fallopian tube and the adipose tissue of the omentum.

During the surgical intervention further 3 units of blood were administered following a decrease in haemoglobin to 73 g/L. The patient returned to the ICU intubated and mechanically ventilated needing haemodynamic support with high doses of norepinephrine (maximum dose 0.4 mcg/kg/min which for the weight of the patient were 28 mcg/min). In view of the high fever registered and the presence of the intra-abdominal fistula, treatment with metronidazole and ceftriaxone was initiated.

Figure 2 Photograph illustrating the right fallopian tube with the origin of the fistula starting from the collateral venous system (arrow on the left side), and the caecum with an orifice (arrow on the right side) indicating the entry of the fistula into the ascending colon.

OUTCOME AND FOLLOW-UP

Following the laparotomy, the patient gradually improved requiring lower doses of norepinephrine and there was no further evidence of bleeding. She was extubated after 7 days. *Bacteroides vulgatus*, was identified in the blood cultures and treatment with metronidazole was completed for a period of 10 days. The patient was discharged from ICU after 9 days. After the acute phase, the patient was followed-up by a hepatologist and she was started on non-cardioselective betablocker, aldosterone antagonist and a loop diuretic. Follow-up esophagogastroduodenoscopy (2 months from the initial hospital admission) excluded the presence of gastro-oesophageal and gastric varices. At the last visit in the hepatology clinic the patient was stable with a compensated Child-Pugh A/B cirrhosis.

DISCUSSION

The pathogenesis of varices is linked to portal hypertension which creates dilatation of the vessels in the portal venous system and an extensive communicating venous network in all of the gastrointestinal system. The site for the formation of gastrointestinal varices is at the gastro-oesophageal junction.⁵

Ectopic varices are dilated portosystemic collateral veins outside the gastro-oesophageal region and the most common sites are the rectum and duodenum, but varices can also be found in the peritoneum, biliary system, urinary bladder, ovaries, vagina and all other abdominal viscera are potential sites of ectopic varices formation.^{7,8}

A potential severe complication of ectopic varices is haemorrhage and only 5% of cases result in bleeding from ectopic varices.⁶⁻⁸

The current case report is an unusual first presentation of complications related to cirrhosis in a patient who claimed to have been in good health prior to this hospitalisation. There are cases described in the literature of patients with cirrhosis who presented with massive vaginal bleeding due to vaginal varices.²⁻⁴

In addition, there have been reports of rare cases of acute small intestinal bleeding due to a portosystemic shunt formation via dilated mesenteric veins and varicose ovarian vein in patients with cirrhosis.^{5,6}

However, to the best of our knowledge, there have been no other cases reported where fallopian tube varices had formed a fistula with the colon leading to massive gastrointestinal bleeding. This case illustrates that this is a rare but possible complication

that could be present in cirrhotic patients. This case shows how ectopic varices can present themselves with a great variation of symptoms, signs and localisation, sometimes even unexpected.

The literature describes cases in which ectopic varices were treated with embolisation and transjugular intrahepatic portosystemic shunt but there are no standardised protocols for the diagnosis and treatment of bleeding caused by ectopic varices, as there are few cases described in the literature.^{11 12} On the other hand as illustrated in this case report massive haemorrhage can occur from ectopic varices and there is limited evidence in the literature to indicate that the use of vasoactive drugs like terlipressin and octreotide could have a benefit in patients with bleeding from ectopic varices.

The description of new cases could help research in the field of ectopic varices, to find better diagnostic and therapeutic strategies in this complex and varied clinical entity.

Learning points

- ▶ Ectopic varices may develop in any of the abdominal viscera and they can have a large spectrum of clinical presentation.
- ▶ Pelvic varices are a rare complication seen in patients with cirrhosis which could lead to massive haemorrhage.
- ▶ Fistula formation between pelvic varices and the gastrointestinal tract should be considered as part of the differential diagnosis in patients in whom this is not identified straight away on radiological investigations.
- ▶ At the moment there are no standardised diagnostic therapeutic protocols for the management of haemorrhage caused by ectopic varices.

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