CASE REPORT

Solid pseudopapillary neoplasm of pancreas: an unusual aetiology for haematochezia

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Accepted 8 June 2018

SUMMARY

Solid pseudopapillary neoplasm of the pancreas is a rare condition. It is mostly seen among young women, with abdominal pain as the presenting complaint. This is a case of a young woman who presented with haematochezia, and was found to have splenic vein thrombosis and left-sided portal hypertension as a result of mass lesion at the tail of the pancreas which was diagnosed as solid pseudopapillary neoplasm.

BACKGROUND

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare condition that occurs predominantly in young women in the second or third decade of life.^{1,2} Due to the rarity of the condition, its natural history remains unclear. The patients may present with an asymptomatic palpable mass or a dull aching abdominal pain. Our patient, however, presented with haematochezia. To the best of our knowledge, this is the first case of SPN causing splenic vein thrombosis and left-sided portal hypertension, thereby resulting in haematochezia due to variceal bleed.

CASE PRESENTATION

A 28-year-old woman presented to the Medicine department with painless bleeding per rectum, three to four episodes (10–20 mL/episode) over the past 2 months. On the morning of presentation, she had passed about 20 mL of blood in her stool. There were no other associated symptoms. She did not have any comorbidity and was not on any regular medications. She did not have any significant family history. On examination, she was overweight (body mass index 28.34 kg/m²). Her vitals and systemic examinations were normal.

INVESTIGATIONS

Her complete blood count showed mild anaemia (haemoglobin 11.4 g/dL). Other investigations like renal and liver functions, electrolytes, Prothrombin time/International Normalised Ratio (PT/INR) and activated partial thromboplastin time (aPTT) were normal. Viral markers (Hepatitis B surface antigen, anti-hepatitis C virus, HIV) were negative. Endoscopic evaluation of the colon showed grade II haemorrhoids (with no red colour signs) at the anal verge, and the visualised colon from rectum to caecum had normal mucosa and vascularity. Ultrasound abdomen showed mild fatty liver and a pancreatic mass. Serum amylase and lipase

levels were mildly elevated (110 U/L and 190 U/L, respectively). Tumour markers like alpha fetoprotein (AFP), carcinoembryonic antigen (CEA), CA 19-9 and CA 125 were within normal limits. The following day, she again had an episode of haematochezia (about 20 mL). Her repeat haemoglobin was 10.5 g/dL. Because the colonoscopy could not fully explain the cause of recurrent bleeding (and as nuclear medicine RBC scan was not available), an upper gastrointestinal (GI) endoscopy was done which showed oesophageal veins and multiple bunches of veins in the fundus of the stomach with red colour signs (figure 1). About 1 mL blood was seen in gastric fundus, but there was no active bleeding. Contrast-enhanced CT of the abdomen revealed a well-defined hypoattenuated pancreatic lesion (30×23×22 mm) with mildly enhancing eccentric mural component in the tail region with dense calcifications at the periphery (figure 2). Superiorly, the lesion was abutting the splenic artery and greater curvature of stomach wall. The rest of the pancreas appeared normal with no ductal dilatation or calcifications. The spleen was mildly enlarged (14 cm). Multiple tortuous splenic and perigastric collaterals were seen at the tail region of the pancreas, and the left gastric veins appeared dilated (figure 3).

TREATMENT

Her operative findings were a 5×4 cm mass lesion at the tail of pancreas. Splenic vein thrombosis was present with dilated collateral veins along short gastric tributaries. The spleen was congested and enlarged. She underwent laparoscopic distal pancreatectomy with splenectomy. Her postoperative period was uneventful. She was discharged on day 7 of admission.

OUTCOME AND FOLLOW-UP

Histological examination of the pancreatic specimen showed a circumscribed neoplasm composed of a monotonous population of cells arranged diffusely and in papillary pattern, with calcified foci of cholesterol clefts; suggestive of SPN. The patient continues to be asymptomatic, with no further episodes of haematochezia, on 4 months review and is on regular follow-up.

DISCUSSION

SPN accounts for only about 0.13%–2.7% of all pancreatic tumours. Females are more prone; and males account for less than 10% of the cases.^{1,2} Sex

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To cite: Aikot S, Manappallil RG, Pokkattil S, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-225332

Figure 1 Multiple bunches of fundal veins in the stomach.

hormones may have a role in the growth or pathogenesis of the tumour.³ They have a low-grade malignant potential and hold an excellent prognosis following complete resection. Clinically, these patients present with abdominal pain or abdominal mass. Anaemia, intestinal obstruction, pancreatitis, bile duct obstruction and haemoperitoneum have also been reported.^{4 5} SPN can occur in any part of the pancreas, but are more common at the tail.⁶ CT imaging show a well-capsulated mass-like lesion with varying solid and cystic components due to haemorrhagic degeneration but lesions <3cm in diameter are purely solid without any cystic component. About 75% of small SPN lack calcification.⁷ Endoscopic ultrasound-guided fine-needle aspiration is a useful diagnostic tool for pancreatic tumours; and about 75% of the SPN cases have been diagnosed preoperatively by this method. However, there is a risk of spread of the tumour.^{8 9} These patients may not show any evidence of pancreatic insufficiency, abnormal liver function tests, cholestasis or elevated pancreatic enzymes. Tumour markers like AFP, CEA,

Figure 2 Contrast CT abdomen showing a mass lesion at the tail of the pancreas. The arrow mark is showing the pancreatic mass lesion.

Figure 3 Contrast CT abdomen (venous phase) showing a splenic vein thrombosis with collateral formation. The circled area is highlighting splenic vein thrombosis.

CA 19-9 and CA 125 are usually within normal range.^{2 10} About 10%–15% of the patients have tumour metastasis at the time of presentation; the common sites being liver, peritoneum, mesentery, omentum and regional lymph nodes.⁵ The differential diagnosis of adrenal mass, pancreatic endocrine tumour, liver cyst or tumour, and pseudocyst pancreas are considered depending on the position of the mass.³ Once the diagnosis of SPN is established, conservative surgical resection with preservation of maximum possible pancreatic tissue is the treatment of choice. Distal pancreatectomy with or without splenectomy, pylorus preserving pancreatoduodenectomy, Whipple operation or enucleation are performed based on the location of the tumour. Portal vein resection is required if there is evidence of tumour invasion. Surgical debulking should be performed in case of metastasis. The prognosis is good, with an overall 5-year survival rate of about 95%.^{2 5} Malignant SPN, also called as solid-pseudopapillary carcinoma, can occur in about 15% of the patients.¹¹ The role of chemotherapy and radiotherapy in SPN

Learning points

- ▶ Haematochezia is present in 15%–20% cases of upper gastrointestinal bleed.
- ▶ Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare condition, primarily affecting young women.
- ▶ Abdominal pain and abdominal mass are the usual complaints. Haematochezia as a presenting feature of SPN in the absence of usual symptoms has not been reported yet.
- ▶ Metastasis is uncommon.
- ▶ Surgical resection of the tumour is the treatment of choice. The role of chemotherapy and radiotherapy is debatable.
- ▶ The prognosis is good, even in the presence of distant metastasis.

is not clear. However, adjuvant chemotherapy and radiotherapy have shown good results in certain unresectable cases.^{12 13} A few cases have also been successfully managed with neoadjuvant chemotherapy or chemoradiotherapy.^{14–16}

GI bleeding can be overt or occult. Overt GI bleed can present as haematemesis, malena or haematochezia. Depending on the site of bleeding, it can be categorised as upper GI, lower GI or obscure (if the site is unknown). Haematochezia is usually a symptom of colonic lesions. However, 15%–20% cases of upper GI bleed can present with haematochezia.¹⁷ This is due to the rapid transit of blood in the bowel before the development of malena and is associated with haemodynamic instability and drop in haemoglobin.

Our patient presented with haematochezia and drop in haemoglobin due to variceal bleed following left gastric portal hypertension as a result of SPN.

Acknowledgements The authors would like to thank Dr George Mathew (Consultant Surgical Gastroenterologist, Baby Memorial Hospital), Dr Anju Chacko (Pathologist, Baby Memorial Hospital) and Dr Dileep Chandrasekhar (Radiologist, Baby Memorial Hospital), Dr Archana Della (Radiologist, Baby Memorial Hospital) for their active participation in patient management.

Contributors SA: review of literature and treating surgical gastroenterologist. RGM: concept, design, review and preparation of manuscript and treating physician. SP: review of literature and treating medical gastroenterologist. AK: resident in-charge.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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