CASE REPORT

Adenomyomatous hyperplasia of the ampulla of Vater presenting as acute pancreatitis

Rafiullah,¹ Sabo Tanimu²

¹Department of Internal Medicine, Saint Clair's Hospital, Weston, Wisconsin, USA

²Department of Gastroenterology, Marshfield Clinic, Weston, Wisconsin, USA

Correspondence to

Dr Rafiullah Khan,
drraziullahkhan@yahoo.com

Accepted 25 January 2014

SUMMARY

We report an interesting and rare case of a man with adenomyomatous hyperplasia of the ampulla of Vater presenting as acute pancreatitis, which to our knowledge, is only the second reported case in the English literature. The patient presented with an acute onset of abdominal pain, nausea and vomiting, without fever, chills or rigours. CT of the abdomen revealed changes of acute pancreatitis with a peripancreatic adenopathy, and abdominal ultrasound revealed a slightly hyperechoic and oedematous head of the pancreas, consistent with acute pancreatitis. Endoscopic retrograde cholangiopancreatography revealed an ampullary lesion. Pathology of the ampullary lesion revealed an inflammatory polyp. Endoscopic ultrasound with endoscopic mucosal resection of the lesion revealed an adenomyomatous hyperplasia. The patient recovered well postendoscopic resection without recurrent pancreatitis or cholestasis.

BACKGROUND

Adenomyomatous hyperplasia and adenomyomas are rare benign lesions of the gastrointestinal tract and hepatobiliary areas. Adenomyomatous hyperplasia of the Vaterian system is extremely rare.¹ The adenomyomas/adenomyomatous hyperplasia of the ampulla of Vater has clinical importance due to its strategic location, unlike in other locations of the gastrointestinal tract that are usually asymptomatic.² Owing to assignments of different names (adenomyomatous hyperplasia, myoepithelial hyperplasia, adenomyoma, adenomyomatosis), it is very difficult to assess the exact incidence of these benign non-neoplastic lesions.² Jutras³ defined the radiological and histopathological features of adenomyomatous hyperplasia, adenomyoma and hyperplastic cholecystoses of the gallbladder in 1960. Kwon *et al*⁴ reported the first case of adenomyoma of the ampulla of Vater presenting as acute recurrent pancreatitis, which is extremely rare. We

report an interesting and rare case of a man, aged 61 years, with adenomyomatous hyperplasia of the ampulla presenting as acute pancreatitis, which to our knowledge, is only the second reported case in the English literature.⁴

CASE PRESENTATION

A 61-year-old man presented with an acute onset of abdominal pain, nausea and vomiting, without fever, chills or rigours. Abdominal examination revealed mild epigastric tenderness. White cell count was $25.5 \times 10^3/\mu\text{L}$ (range $4.1\text{--}10.9 \times 10^3/\mu\text{L}$) with 88% neutrophils (range 34–67%); haemoglobin was within normal range at 14.4 g/dL. Liver function test revealed a normal bilirubin of 1.3; however, aspartate aminotransferase was elevated at 190 U/L (range 15–46 U/L), alanine aminotransferase was elevated at 169 (range 10–61 U/L), and an alkaline phosphatase was elevated at 147 U/L (range 40–125 U/L). Amylase was significantly elevated at 1855 U/L (range 25–100 U/L), and lipase was greater than 285 U/L (range 13–50 U/L).

A CT scan of the abdomen revealed changes of acute pancreatitis with a peripancreatic adenopathy. The common bile duct was dilated to 10 mm with abrupt discontinuation at the head of the pancreas on the background of a mildly dilated pancreatic duct measuring 4 mm at the head of the pancreas, which was concerning for a stone or periampullary neoplasia (figure 1). The gallbladder revealed no evidence of a stone.

Abdominal ultrasound revealed a normal gallbladder with a common bile duct of 8 mm without a stone and a slightly hyperechoic and oedematous head of the pancreas, with a pancreatic duct measuring 4 mm at the body of the pancreas, consistent with acute pancreatitis (figure 2). Endoscopic ultrasound revealed a 2.4×2.1 cm multilobulated hyperechoic ampullary density with a semipedunculated base occupying 25–50% of the superior circumference of the ampulla, with a valve-like effect

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To cite: Rafiullah, Tanimu S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203151

Figure 1 (A) Ampullary adenomatous hyperplasia with an indwelling biliary stent (B).

Figure 2 The common bile duct and pancreatic duct were uninvolved and separate from the lower half of the ampulla.

on the pancreaticobiliary orifice. There was an indistinct margin with the head of the pancreas because of residual peripancreatic stranding. The pancreatic duct measured 4 mm in the head of the pancreas with progressive diminution to the tail of the pancreas. The common bile duct measured 1.1 cm with progressive tapering to the ampulla. There were two peripancreatic adenopathies, measuring 2.1×1.1 cm and 7.8×3.8 mm, that were reactive by fine-needle aspiration cytology. Endoscopic retrograde cholangiopancreatography (ERCP) revealed an ampullary lesion with a valve-like effect on the biliary and pancreatic orifices (figure 3). A 7 Fr $\times$ 7 cm pigtail biliary stent was placed across the ampulla into a dilated common bile duct at 1.1 cm. The pancreatic duct was intentionally not cannulated.

Pathology of the ampullary lesion revealed an inflammatory polyp versus inflammatory changes overlying an unsampled neoplastic lesion (figure 4). Endoscopic ultrasound with endoscopic mucosal resection of the lesion revealed an adenomyomatous hyperplasia. The patient recovered well postendoscopic resection without recurrent pancreatitis or cholestasis.

DISCUSSION

Adenomyoma/adenomyomatous hyperplasia is defined as a nodular lesion composed of smooth muscle and epithelial cells.^{2 5} Adenomyomas/adenomyomatous hyperplasia is extremely rare, and by 2003 only 33 cases had been reported in the English literature.² The most common location is the gallbladder; however, adenomyoma has also been reported in the stomach, small bowel, extrahepatic bile duct and ampulla of

Vater.^{1 2 4 5} In one case series of 13 patients reported by Handra-Luca *et al*² from France, the average age of presentation was 63 years (ranging from 38 to 78 years), consistent with the age of our patient at 61 years. In the same case series, the male-to-female ratio was 6:7.² The clinical presentation is usually variable depending on the location of the lesion. It can be asymptomatic or may present with jaundice, epigastric pain, right upper quadrant pain or non-icteric cholangitis.² Our patient presented with acute abdominal pain associated with nausea, vomiting, cholestasis and pancreatitis, secondary to adenomyomatous hyperplasia of ampulla of Vater, which to our knowledge, is only the second reported case in the literature.⁴

Adenomyomas are histologically differentiated from chronic papillitis (associated with benign conditions such as gallstones and duodenal inflammation) by the absence of fibrosis.⁶ The histogenesis of adenomyomatous hyperplasia is not clear. The hypothesis describing these lesions as incomplete heterotrophic pancreas is the most widely accepted,⁷ and can explain the presence of intestinal glands and Brunner's glands within the same lesion.⁸ Narita and Yokoyama⁶ believe these lesions may be inflammatory in origin.⁹

The preoperative and intraoperative diagnosis can be challenging, and patients are frequently misdiagnosed as having ampullary neoplasms, leading to extensive surgery such as pancreaticoduodenectomy.² Our patient was also initially diagnosed with ampullary adenoma on histopathological examination, which was later revised to adenomyomatous hyperplasia. The newer immunohistochemical markers, like low proliferative activity (Ki67), cytokeratin 7 expression in epithelial cells and absence of cytokeratin 20 expression help in differentiating adenomyoma/adenomatous hyperplasia from ampullary neoplasm. This will help in the decision of endoscopic resections and limited surgical options in the treatment of these benign lesions.² There are no specific guidelines for treatment of adenomyoma/adenomyomatous hyperplasia. The diagnosis of ampullary lesions (including ampullary tumours and benign lesions such as adenomyomatous hyperplasia) is made using a combination of diagnostic endoscopy, imaging and histopathology. The primary goal is ruling out any occult malignancy or tumour, that can only be carried out with histopathological examination of the completely resected mass^{9–11} (as in our case). Imaging studies most commonly performed are abdominal ultrasound, CT, MR cholangiopancreatography, and percutaneous transhepatic cholangiography.

Ampullary lesions can be detected by ERCP in almost all patients; however, biopsy via ERCP cannot exclude occult

Figure 3 En bloc ampulectomy with resection of the right hemicircumference of the ampulla.

Figure 4 The resected specimen showing the submucosal layer.

ampullary carcinoma (in our case, it was falsely negative for ampullary adenomyomatous hyperplasia).^{10–14} The false-negative rate for endoscopic biopsy is between 16% and 60%. The false-negative rate can be decreased to almost zero by performing the biopsy 2–10 days after sphincterotomy.^{12 15 16} The potential roles of intraductal ultrasound, magnifying endoscopy, capsule endoscopy and narrow-band imaging endoscopy in diagnosing ampullary adenoma are still under investigation.^{17–20} Patients with ampullary lesions (including ampullary tumours and benign ampullary lesions, depending on the size and preoperative/intraoperative diagnosis) have three treatment options. Options include local surgical resection or pancreaticoduodenectomy, which is the traditional surgical approach.²¹ Our patient underwent endoscopic mucosal resection, and his subsequent pathology report confirmed the diagnosis of adenomyomatous hyperplasia.

Learning points

- ▶ Adenomyomatous hyperplasia is rare but should be considered in the differential diagnosis when evaluating ampullary lesions.
- ▶ Adenomyomas are generally considered benign lesions, but the possibility of malignant potential cannot be completely ruled out.
- ▶ Occult malignancy in the ampullary lesions can only be excluded by histopathological examinations of the completely resected lesions.
- ▶ The preoperative and intraoperative diagnosis of this benign non-neoplastic lesion can avoid extensive surgical interventions, like pancreaticoduodenectomy.

Acknowledgements We thank Dr Anil babu Thogarucheti and Ms Debra Knipple Librarian for helping us in the literature search. We also thanks Ms Marie Fleisner of Marshfield Clinic Research foundation editorial specialist for editing this manuscript.

Contributors All authors have participated in the planning, execution or analysis of the study. All authors affirm that the final manuscript has been seen and approved by them for submission to BMJ.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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