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CASE REPORT

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Myoepithelial hamartoma of the duodenal wall

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Abstract A rare case of myoepithelial hamartoma of the duodenal wall is presented, and previous case reports found in the literature are reviewed. Myoepithelial hamartomas are thought to arise from displaced pancreatic anlage present along the gastrointestinal tract during embryogenesis, which can differentiate into various pancreatic elements; the most highly differentiated form is heterotopic pancreas. An alternative theory is pancreatic metaplasia of endodermal tissues. We describe a 41-year-old man who presented with abdominal pain and vomiting. CT scanning revealed a mass at the head of the pancreas. A pancreaticoduodenectomy was performed for presumed cystadenoma. Histology of the mass revealed a disorderly arrangement of smooth muscle, dilated and nondilated ducts, pancreatic acinar tissue and mucus glands. The relationship of myoepithelial hamartomas involving the small bowel to similar lesions in the stomach, bile ducts and gallbladder is discussed.

Key words Hamartoma · Adenomyoma · Pancreas · Choristoma · Small intestine

Introduction

Myoepithelial hamartomas in the gastrointestinal tract arise during embryogenesis from pancreatic metaplasia or, alternatively, from a displaced pancreatic anlage [10]. The anlage is capable of differentiation into normal pancreas, which is termed heterotopic pancreas, or into various abnormally arranged pancreatic elements. The latter lesions may show only pancreaticobiliary ducts surrounded by smooth muscle, or may in addition contain a disordered arrangement of pancreatic acini and mucus glands. Because of the histological variability, these less highly differentiated forms have been referred to by various names, including incompletely differentiated pancreas, adenomyomas, adenomyosis, and foregut choristomas. In this paper, 'myoepithelial hamartomas' will be used to refer to this spectrum of lesions, whereas only in those lesions with normally arranged pancreas will the term 'heterotopic pancreas' be used.

Myoepithelial hamartomas occur most commonly in the stomach and small bowel. Thirty-eight cases have been reported in the stomach, making it the commonest location [15]. Eighty-five percent of such lesions are located in the antrum and 15%, in the pylorus. Approximately one-third contain pancreatic acini and/or mucus glands. The small bowel is the second most frequent site, with 14 cases reported in the English literature. We report the case of a 41-year-old man who underwent a pancreaticoduodenectomy for a mass lesion at the head of the pancreas when fine-needle aspiration suggested cystadenoma.

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Clinical history

A 41-year-old man with a long-standing history of alcohol abuse and a recent diagnosis of hepatitis C infection was referred to our institution complaining of fatigue, malaise, weight loss and intermittent episodes of severe mid-epigastric pain, and nausea and vomiting over the past 2–3 days. He had experienced similar episodes of nausea and vomiting several times a month over the past 6 years, frequently after periods of heavy drinking. These episodes lasted for 1–2 days. The only serum amylase check made during this period was on 31 August 1995; the level was minimally elevated at 135 U/l (normal, 20–112 U/l). No serum lipases were measured. Over the past 6 months, these episodes had become more frequent and severe with recent onset of increased weakness and weight loss. Physical examination showed no stigmata of chronic liver disease. Abdominal examination revealed nothing significant except mild mid-epigastric tenderness without back pain.

Laboratory studies showed an elevated white count of 14,000 mm^3 with a normal differential. Serum lipase was 356 U/l (normal, 50–250 U/l), and amylase was normal. Liver function tests and enzymes were also normal as were serum electrolytes. CT examination of the abdomen with contrast revealed a large complex mass (7 × 4.4 × 2.5 cm) of cystic and solid components

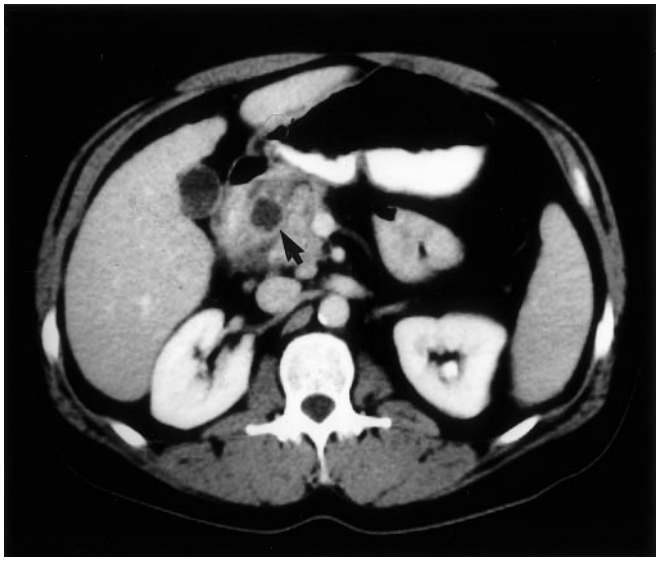

Fig. 1 CT of abdomen showing mass in the duodenal wall (*arrow*)

arising between the duodenum and pancreas (Fig. 1). There was evidence of lymphadenopathy in gastrohepatic, celiac and pre-caval spaces. There was no evidence of duodenal obstruction. Liver, pancreas and spleen were otherwise normal. Endoscopic examination demonstrated a submucosal mass adjacent to the ampulla of Vater. Biopsy of the duodenal mucosa showed mild nonspecific duodenitis with reactive lymphoid nodules, prominent Brunner's glands and focal gastric foveolar metaplasia without evidence of neoplasia. CT-guided needle aspiration of the mass was performed: the aspirates contained hyperplastic duct epithelium, histiocytes and mucus suggestive of mucinous cystadenoma without atypia.

The patient underwent pancreaticoduodenectomy for a mass at the head of the pancreas presumed to be cystadenoma. At laparotomy, the patient was found to have a large bulky mass in the periampullary region with no evidence of peritoneal involvement. The mass could be palpated in the head of the pancreas. The tail and body of the pancreas were soft, and none of the adjacent vasculature was involved. The mass was resected at the level of the pancreatic neck and the specimen sent to pathology along with a large peripancreatic lymph node. The procedure was completed without complications, and the patient's postoperative course was uneventful. He was discharged on postoperative day 22 in good condition.

Pathological findings

The resected specimen consisted of distal pylorus, duodenum and a portion of pancreas. The pylorus was unremarkable, and the duodenal mucosa proximal to the ampulla of Vater was flattened and granular. The ampulla was prominent. The periampullary duodenal wall was expanded by an ill-defined, white-tan mass measuring 6 × 3 cm, which extended into the adjacent pancreas and surrounded the distal common bile duct. Sectioning revealed a slightly trabeculated cut surface with pinpoints and microcysts measuring 0.5 cm in greatest circumference. The appearance was reminiscent of uterine adenomyosis, where the glandular epithelium is slightly more tan than the surrounding white-grey trabeculae of smooth muscle and protrudes slightly from the cut sur-

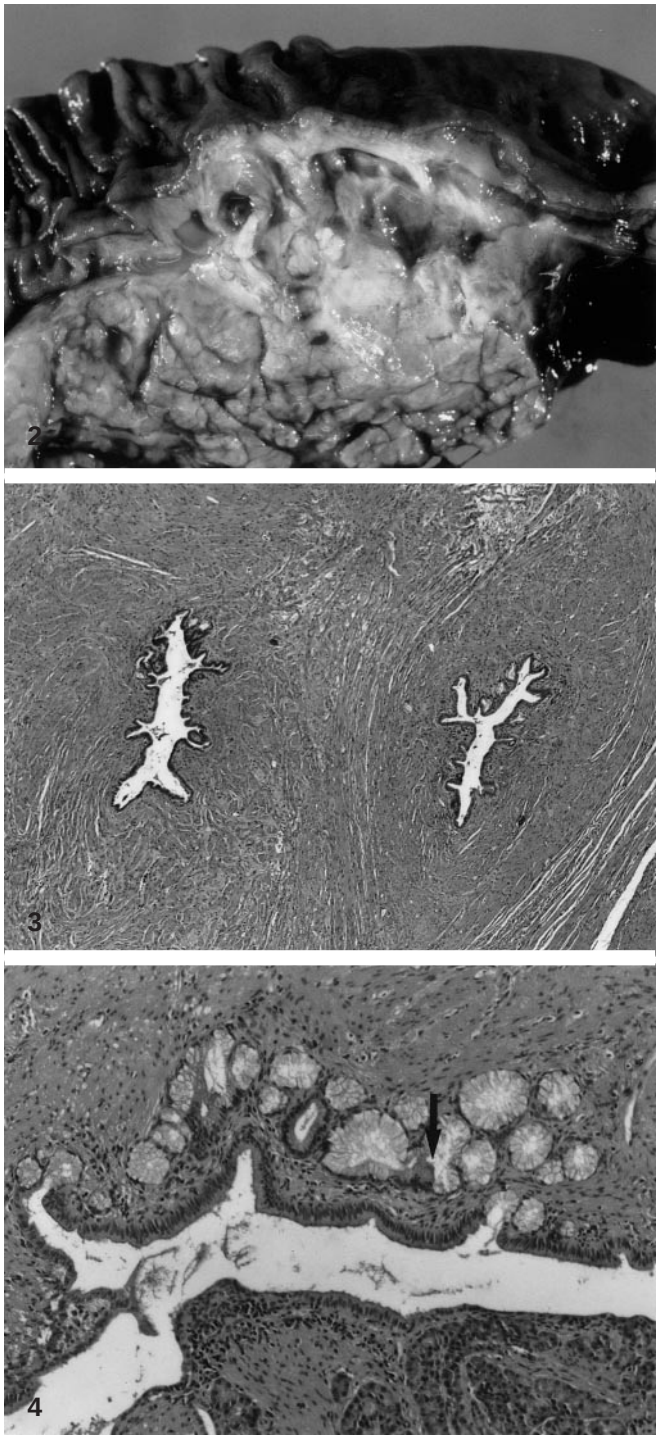

Fig. 2 Gross photograph showing diffuse thickening of the duodenal wall

Fig. 3 Microscopically the lesion consists of dilated ducts surrounded by whorls of smooth muscle. H&E, ×20

Fig. 4 In some areas pancreatic acini (*lower right* part of the field) and subepithelial mucus glands are seen. Areas of transition between ductal epithelium and mucus gland epithelium are seen (*arrow*). H&E, ×100

Table 1 Cases of myoepithelial hamartoma involving the small bowel in the English literature (*ns* not stated)

Case no.	Reference	Age	Location	Size (cm)	Presenting symptoms
1	[3]	64 years/M	Jejunum	1.0	Incidental finding at autopsy
2	[3]	46 years/F	Duodenum	ns	Diarrhoea, fever and abdominal pain
3	[12]	2 days/F	Ileum	ns	Bowel obstruction
4	[1]	54 years/M	Duodenum	4.0	Common and pancreatic duct obstruction
5	[4]	82 years/F	Ileum	2.0	Intussusception
6	[9]	74 years/F	Ileum	4.0	Intussusception
7	[5]	9 months/M	Ileum	1.2	Intussusception
8	[5]	79 years/M	Ileum	0.6	Incidental finding
9	[11]	33 years/M	Duodenum	4.0	Weight loss, vomiting
10	[2]	5 months/F	Ileum	0.8	Intussusception
11	[2]	3 years/M	Ileum	0.8	Nausea, vomiting and abdominal pain
12	[6]	2 years/M	Ileum	1.4	Rectal bleeding
13	[13]	44 years/M	Ileum	2.0	Intussusception
14	Present case	41 years/M	Duodenum	6.0	Nausea, vomiting and abdominal pain

face. No areas of haemorrhage or necrosis were identified (Fig. 2). Histologically, the mass consisted of irregularly arranged whorls of smooth muscle surrounding ductal structures (Fig. 3). A proliferation of nerve tissue accompanied the smooth muscle proliferation. The ductal structures were lined by columnar cells with basally arranged uniform nuclei. No cytological atypia, goblet cells or argentaffin cells were identified. Many of the ducts were dilated, and in these the epithelium was cuboidal and their lumina contained inspissated material. Small nests of pancreatic acini were also present adjacent to the ductal structures in some areas, together with mucus glands just beneath the epithelium of the ducts. In several areas, a transition from duct epithelium to mucus gland epithelium was noted in the same structure (Fig. 4). No pancreatic islets were identified. The lesion extended from just below the muscularis mucosa to the adjacent pancreas. The overlying duodenal mucosa showed reactive changes consisting of mildly flattened villi and mild nonspecific duodenitis. Prominent Brunner's glands are also noted in the submucosa. The pancreas was histologically unremarkable.

Immunohistochemically, NSE, synaptophysin and S100 outlined the proliferation of nerves within the smooth muscle bundles. Interestingly, the synaptophysin and NSE also outlined aggregates of cells that resembled islets also scattered randomly within the smooth muscle bundles. These aggregates were not initially appreciated on H&E staining. Their islet cell nature was confirmed with positive staining for both glucagon and insulin. Keratin stain Cam 5.2 stained the bile duct epithelium, acinar and islet tissue. Staining with CEA stained both the duct cells and, focally, the acinar cells. Staining with amylase outlined the acinar cells.

Discussion

Including this report, 14 cases of myoepithelial hamartoma involving the small bowel have been reported with histological descriptions in the English literature (Table 1). Cases of heterotopic pancreas have been excluded. The patients' ages range from 2 days to 82 years; 10 of the cases occurred in male and 4 in female patients. The most common presentation is intussusception and intestinal or biliary obstruction, depending on the location of the lesion. In 2 cases, the lesion was an incidental finding. Of the fourteen cases, 9 have occurred in the ileum, 4 in the duodenum, and 1 in the jejunum. The lesions range in size from 0.6 cm to 6.0 cm in greatest dimension. They tend to be well circumscribed and involve the submucosa, with some extending into the muscularis propia. Only 1 other case containing pancreatic acini has been described [3]. Several reports have noted squamous metaplasia and the presence of goblet cells and argentaffin cells within the ductal epithelium.

Gallbladder lesions referred to as adenomyomas or adenomyosis are not pathogenetically related to this lesion. Although their histology shows epithelial structures and smooth muscle, such lesions are believed to arise from diverticular disease of the gallbladder and are not related to heterotopic pancreas. Lesions occurring in the bile ducts have also been referred to as adenomyomas. Some involve the ampulla of Vater [7, 14] and resemble these lesions histologically. Those described in the larger bile ducts [8] differ histologically in that larger ductal structures are surrounded by smaller ductal structures. It is not clear whether these are pathogenetically related.

Anything that can occur in the pancreas can happen in heterotopic pancreatic tissue, including pancreatitis and carcinoma. It is important to recognize the existence of this rare entity in the small bowel, because clinically it can mimic neoplasia and it may occasionally harbour a carcinoma.

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