Pancreatic Heterotopia in Jejunum presenting as small Bowel Obstruction

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Abstract

Heterotopic pancreas is a congenital malformation, defined by the presence of pancreatic tissue outside the usual anatomical location of the pancreas. Most patients are asymptomatic and when present, symptoms are nonspecific. Rarely, they become evident when associated with complications such as inflammation and obstruction. A 48- year old male presented with features of small bowel obstruction. A submucosal polyp was detected in the jejunum intraoperatively and excised. Diagnosis of heterotopic pancreas was made on histopathological examination.Though rare,heterotopic pancreas should be considered in the differential diagnosis of small bowel obstructionand of submucosal polyps in the small intestine. Pre-operative diagnosis is difficult. Histopathological examination is mandatory for confirmation. Frozen sections may help in distinguishing these lesions from malignancies and gastrointestinal stromal tumors. Jejunum is an unusual site for heterotopic pancreas. It rarely presents as small bowel obstruction.We present this case to highlight its rarity, clinical manifestations, diagnostic tools available, pathogenesis and histopathology.

Key words : Heterotopic pancreas, jejunum, obstruction, histopathology

Introduction

Heterotopic pancreas refers to the presence of pancreatic tissue outside its normal location. It lacks anatomic or vascular relation to orthotopic pancreas. The term was first used by de Castro et al[1]. It is a relatively infrequent lesion, with varying incidence in different clinical and autopsy studies [2]. Typically, it is detected as an incidental finding at laparotomy or autopsy anywhere along the gastrointestinal tract[3]. Most common site is the stomach. Jejunum is a rare location for heterotopic pancreas [4]. Histologically, it contains an admixture of tissues found in the normal pancreas. Symptoms, when present depend on the location and local effects of pancreatic tissue. Complications include inflammation, obstruction and malignant transformation [5].

Case report

A 48-year old male presented with vomiting and pain abdomen. A clinical diagnosis of gastric outlet obstruction was made. Endoscopy showed an ulcer measuring 3x2cms in the first part of the duodenum. The scope could not be negotiated beyond the second part of the duodenum. Truncal vagotomy with gastrojejunostomy was planned. During surgery, a polyp measuring 2x1cm was found in the proximal jejunum. The lesion was excised and sent for HPE. Symptoms resolved within a fortnight.

Gross Pathology

A grey white to grey brown polypoid mass measuring 1.0cm across, partly covered with mucosa was seen. Cut surface was yellowish in colour with a lobulated appearance.

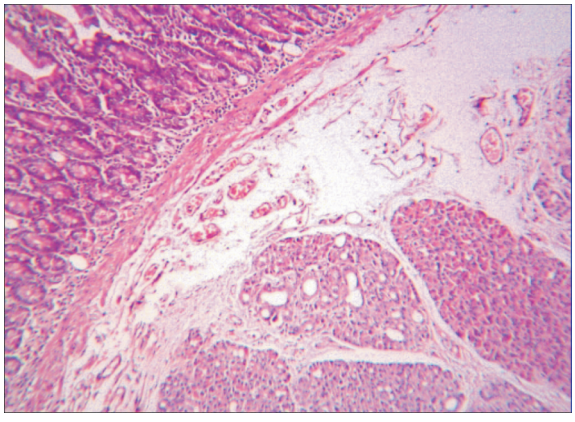

Figure1. Photomicrograph showing jejunal mucosa with benign pancreatic tissue in the submucosa (H& E, 10x)

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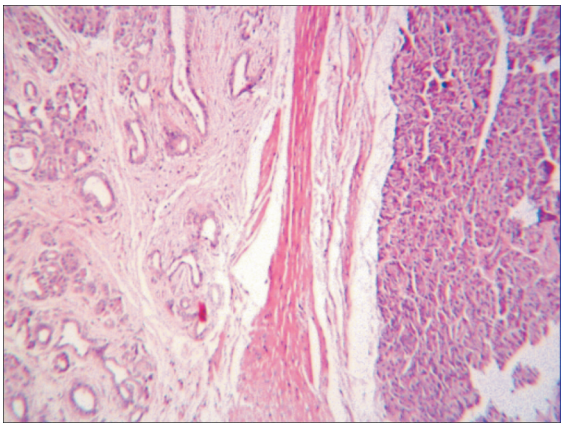

Figure 2. Photomicrograph showing lobules of pancreatic acini and ducts surrounded by fibromuscular stroma (H&E,10x)

Microscopy showed intact mucosa of jejunum (Figure1). The submucosa showed well-circumscribed lobules of benign pancreatic acini interspersed with ducts and surrounded by fibrous stroma (Heinrich type II) (Figures 2 and 3). The heterotopic pancreatic tissue was seen involving the muscular layer of the jejunum (Figure 4).

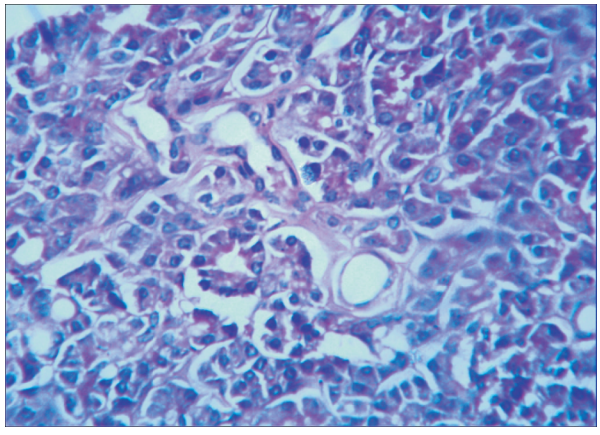

Figure 3. Photomicrograph showing benign pancreaticini and ducts (H&E,40x)

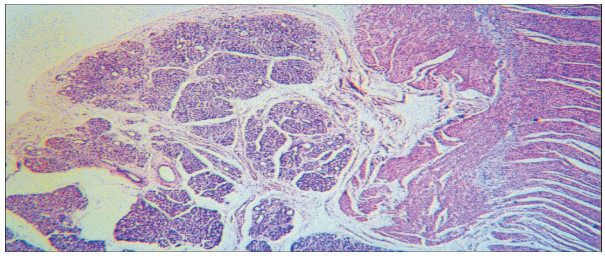

Figure 4. Photomicrograph showing lobules of benign pancreatic tissue involving the muscularis propria of the jejunum(H&E,10X)

Discussion

Heterotopic pancreas is relatively rare. It is defined as pancreatic tissue that lacks anatomic and vascular communication with the normal pancreas, possessing its own duct system and blood supply. Jean-Schultz was the first to describe this congenital abnormality [6]. The pathogenesis of this entity is not known. The normal pancreas is derived from the dorsal and ventral 'anlagen'.These primitive foregut buds fuse to form a single gland during rotation of the foregut [7]. Heterotopias are believed to arise either by misplacement of pancreatic fragments during rotation of the foregut or from metaplasia of endodermal tissue in the submucosa during embryologic development of the gastrointestinal tract [8].

The precise incidence of heterotopic pancreas is not known; rates range from 0.1% to 0.2% in various autopsy studies [6]. The clinical prevalence is estimated to be one in every five hundred upper gastrointestinal surgeries [8]. It can be located anywhere in the gastrointestinal tract, most commonly in the stomach.Jejunum is a rare location for ectopic pancreas [4]. Other unusual locations include spleen, liver, mesentery, skin and lymph nodes.

Although congenital in origin, heterotopic pancreas usually manifests in adults, commonly in the sixth decade of life. It is possible that the ectopic tissue may reach significant size with age to produce symptoms in adult life. Most patients are asymptomatic. Symptoms when present vary depending on the location and size of the lesion. Common symptoms include upper abdominal pain, nausea, vomiting and gastrointestinal bleeding. Symptoms are related to local secretion of hormones and enzymes resulting in irritation, inflammation and spasm[3,9]. These features may resemble carcinoid syndrome. When the muscular layer is involved, the heterotopic tissue may act as a foreign body, resulting in local spasm, dysmotility and persistent vomiting, as experienced by our patient. Small bowel obstruction from heterotopic pancreas is rare. Intestinal obstruction is usually associated with intussusception[10]. The obstruction in the present case was not associated with intussusception. Symptoms resolved within a fortnight following surgery in the index case.

Preoperative diagnosis of heterotopic pancreas is difficult despite advances in diagnostic techniques. Radiologic features include a characteristic well defined, dome-shaped filling defect with central umblication. On CT with intravenous contrast, most heterotopias enhance brightly similar to orthotopic pancreas. Inflammation tends to reduce the enhancement, making these findings nonspecific. On endoscopy, it appears as a broad based, umblicated submucosal lesion; it is difficult to distinguish them from gastrointestinal stromal tumors. No endoscopic correlation was available in the present case as the site of lesion was inaccessible due to local spasm. Capsule endoscopy and single bowel enteroscopy can be used to visualise inaccessible small bowel lesions. Single bowel enteroscopy also facilitates biopsies, although they are usually superficial and inadequate.Endoscopic ultrasound is the investigation of choice for evaluating submucosal lesions in the gastrointestinal tract. Endoscopic ultrasound with fine needle aspiration has a high degree of sensitivity[3]. Frozen sections are helpful in differentiating primary from metastatic pancreatic tissue and to exclude stromal tumors[6]. Histopathological examination is mandatory for confirmation. In the index case too, following excision, the diagnosis was established on histopathological examination.

Grossly, heterotopic pancreas appears as a submucosal polyp or an intramural mass. Cut section resembles normal pancreatic parenchyma, with a lobulated appearance, yellow colour and an average size of 2.0 to 4.0 cm. The excised polyp in the present case was submucosal in location and measured 1 cm across. The external surfacewas covered with mucosa. Cut surface had a lobulated, yellowish appearance.

Klob was the first to describe the histological appearance of heterotopic pancreas. It may contain any admixture of tissues found in the normal pancreas. Heinrich classified heterotopic pancreas into three types [8].

Type I: ducts, acini and endocrine islets;
Type II: ducts and acini;
Type III: ducts with few acini or dilated ducts only, so called adenomyoma.

Others divide them into four subtypes [2,3].

Type I - comprising all cell types - total heterotopia;
Type II - comprising ducts only - canalicular heterotopia;
Type III - comprising acinar cells only - exocrine heterotopia;
Type IV - comprising islet cells only - endocrine heterotopia.

Microscopic examination of the excised tissue in the present case showed intact mucosa of the jejunum. The submucosa showed well-circumscribed lobules of benign pancreatic acini interspersed with ducts and surrounded by fibrous stroma. The heterotopic tissueinvolved the muscularis propria of the jejunum. This was classified as type II heterotopic pancreas, Heinrich classification.

Although rare, pancreatitis, islet cell tumour, and adenocarcinoma are some of the complications that may arise in ectopic pancreatic tissue[5]. Pancreatic tissue, in the present case showed normal morphology. Surgery is the treatment of choice in symptomatic patients. The lesion has to be excised when incidentally detected on the operating table, during surgery for other causes, as was the case in our patient. The literature is equivocal on the management of asymptomatic, incidental lesions. Some authors are of the opinion that benign asymptomatic lesions do not require surgical intervention. It must be stressed that clinching a histologic diagnosis is a prerequisite. In view of the difficulty in diagnosing heterotopia even with newer modalities like capsule endoscopy and single bowel enteroscopy, surgical resection may be the only option in most cases.

Conclusion

Heterotopic pancreas is relatively rare andmost cases remain clinically silent. It must be considered in the differential diagnosis of small bowel obstruction. Preoperative diagnosis is difficult. Most investigations are nonspecific. It is interpreted as the more common epithelial or mesenchymal polyp resembling stromal tumour, on endoscopy. Frozen section is recommended for submucosal exophytic masses. Histologic confirmation is a must to exclude malignancy.

References

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