

Original Article

Gastric pancreatic heterotopia: A rare disease entity

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Abstract

Introduction: Heterotopic pancreas, a lesion that is rarely encountered, can be located anywhere within the gastrointestinal tract. It is commonly asymptomatic though most patients may present with pain. Its submucosal location makes preoperative diagnosis difficult. This study aims to report a small series of cases of this disease which has a very low incidence and to enumerate its clinical and endoscopic features.

Methods: From the three year archives of the histopathology department of Kathmandu Pathlab and Diagnostic Centre, all cases reported as heterotopic pancreas were collected from Jan 2013-Dec 2016. Prevalent details were extracted from the requisition forms.

Results: Out of 3,000 samples received, five cases were diagnosed as having heterotopic pancreas. The age range for the disease was 20-50 years with a male predominance. Mean diameter of the lesion was 1.6cm. The endoscopic diagnosis was heterotopic pancreas in one case whereas for others it was gastrointestinal stromal tumor (GIST). Endoscopic biopsies were non diagnostic in all cases and the diagnosis was made on local surgical resection specimen. Pain was the common presenting symptom. Microscopy showed Type 1 lesions in all cases with involvement of the muscularis propria in one case. Other four cases had involvement upto submucosa.

Conclusion: Though a rare entity, pancreatic heterotopias should be considered in the differential diagnosis of submucosal gastrointestinal lesion. The diagnosis of ectopic pancreas is still challenging in spite of the improvement and development in the diagnostic modalities. Symptomatic relief is provided by surgical resection and surgery is recommended particularly if diagnostic dilemma exists.

Keywords: endoscopy, heterotopic pancreas, histopathology

Introduction

Heterotopic pancreas, a lesion that is rarely encountered, can be located anywhere within the gastrointestinal tract.

¹ It is commonly asymptomatic though most patients may present with pain. Due to its submucosal location preoperative diagnosis is difficult. This study aims to report a small series of cases of this disease which has very low incidence and to enumerate its clinical and radiological features.

Methods

From the three year archives of the histopathology department of Kathmandu Pathlab and Diagnostic

Centre, all cases reported as heterotopic pancreas were collected from Jan 2013-Dec 2016. Prevalent details were extracted from the requisition forms.

Results

Out of 3,000 samples received in the histopathology department in Kathmandu Pathlab and Diagnostic Center, five cases were diagnosed as having heterotopic pancreas. The age range for the disease was 20-50 years with a male predominance. Mean diameter of the lesion in these cases was 1.6cm. The endoscopic diagnosis was heterotopic pancreas in one case whereas for others it was gastrointestinal stromal tumor (GIST).

Endoscopic biopsies were non diagnostic in all cases and the diagnosis was made on local surgical resection specimen. Table 1 displays the characteristics of all cases.

Discussion

By definition, pancreatic heterotopia is a pancreatic tissue lying outside its normal location without anatomic or vascular connection with pancreas.¹ In 1927, it was first described as an ileal diverticulum.² Though it can be located in any part of the gastrointestinal tract, it is commonly encountered in the stomach (25-38%), duodenum (17-36%) and jejunum (15-21%). Rarely, it has been seen in the esophagus, biliary tract gall bladder and mesentery.³ It has also been reported in spleen, mediastinum and fallopian tubes.¹ 85-95% of gastric heterotopias are located in the antrum mostly along the greater curvature, either on the anterior or posterior wall.⁴ All five cases in this series were located in the gastric antrum along the greater curvature in the anterior wall.

At autopsy, the incidence of heterotopic pancreas was reported as 0.11-0.21% with a male predominance (M: F is 3:1).⁵ The age range at which it is seen is 30-50 years.⁵ In this study heterotopic pancreas was seen in the age range of 20-50 years and mainly it was found in males as with other studies.

The maximum diameter of heterotopic pancreas described in literature is 1-2cm.⁶ The mean diameter in this study was 1.6cm. The symptom produced by the lesion depends on the relative size of the lesion.

Pancreatic ectopic tissue may present as abdominal pain and distension, although mostly it is silent and asymptomatic. Similar changes as that seen in the normal pancreas including pancreatitis, islet cell tumor, pancreatic carcinoma and pancreatic cyst can also be seen in these ectopic pancreatic tissue.⁶ In some cases of heterotopic pancreas, malignant transformation has been reported.⁷⁻⁸ For the histopathological diagnosis of malignancy in heterotopic pancreas, the malignant tissue should be within or in close proximity to the ectopic focus and a transition should be seen between the carcinomatous and the normal pancreatic tissue.¹ Fully developed acini and ducts should also be present in the non-neoplastic ectopic pancreatic rest. The prognosis of adenocarcinoma arising in an ectopic pancreas is found to better than adenocarcinomas arising in normally located pancreatic tissue.¹

Different theories have been presented about the pathogenesis of heterotopic pancreas. The most accepted theory states that during the development of normal pancreas from several evaginations that arise from the wall of the duodenum, one or more evagination remains in the bowel. The migration of this embryonic remnant tissue along with the development of the gastrointestinal tract leads to the ectopic pancreatic tissue.⁴ Other theories evolved state that, pancreatic metaplasia of the endodermal tissue located in the gastric submucosa may occur during embryogenesis which leads to ectopic pancreatic tissue.⁴

Endoscopy in heterotopic pancreas typically reveals a firm; round to oval submucosal lesion with central depression which corresponds to the opening of a duct. This gross feature at endoscopy of a lesion with a central dimple or umblication favors a provisional diagnosis of heterotopic pancreas.⁹ This typical finding was endoscopically found in one case in this study. Since such finding was absent in other cases, in the presence of a submucosal lesion provisional diagnosis of GIST (gastrointestinal stromal tumor) was made at endoscopy.

Literature states that such typical endoscopic finding may not be seen in all cases.¹⁰ Such lesions lacking a typical endoscopic findings can be diagnosed by a new diagnostic modality called endoscopic ultrasonography (EUS). In EUS, if a subepithelial lesion appears as an inhomogeneously hypoechoic mass in the third and fourth layers, ectopic pancreatic tissue should be considered in the differential diagnosis.¹⁰ Other features described which suggests an increased possibility of heterotopic pancreas are; indistinct borders, lobulated margins and/ or presence of anechoic duct like structure.¹⁰ EUS is a useful adjunctive tool in detecting ectopic submucosal rest with size ranging from 0.5 to 2 cm. In all five cases in this study EUS was not done.

In this study, all cases microscopically showed ducts and acini (Fig 1). Four cases (80%) had involvement of the submucosa whereas a single case showed involvement of the muscularis propria as well (Fig 2). Studies show involvement of submucosal layer in 73%, muscularis in 17% and subserosal layer in 10% of cases.¹¹

The histopathological classification of heterotopic pancreas was proposed by Heinrich in 1909 which was further modified by Gaspar- Fuentes in 1973.¹ According to the classification : Type I is typical pancreatic tissue

with acini, ducts and islet cells similar to that of normal pancreas. Type II comprises pancreatic ducts only referred to as canalicular variety. Type III consists of acinar tissue only referred to as exocrine pancreas. Type IV is characterized by islet cells only referred as endocrine pancreas.¹²

All five cases presented with abdominal pain and discomfort. Studies have shown that pain is a common presenting symptom in heterotopic pancreas.¹³ Pain may be caused by the exocrine and endocrine function of the ectopic pancreatic tissue which secretes hormones and enzymes that is responsible for inflammation and/or clinical irritation of the involved tissue.¹³ Erosion of mucosa may lead to hemorrhage, ulceration or perforation especially if the pancreatic tissue rests in the small intestine.¹³ In pre-pyloric ectopic pancreatic rest, symptoms of gastric outlet obstruction can be seen while obstructive jaundice is the presenting symptom in pancreatic heterotopia of the bile duct. These are the symptoms relating to the mass effect of the lesion. Size larger than 1.5cm in maximum dimension is likely to produce symptoms.¹³ Jejunal lesions can rarely be the cause of intestinal obstruction and intussusception.¹⁴⁻¹⁶

Since endoscopic biopsies are superficial, heterotopic pancreas which is a sub mucosal lesion will yield a non diagnostic endoscopic biopsy result in majority of the cases.¹⁷ Literature has described the combination of EUS along with FNA (fine needle aspiration) of the sub mucosal gastrointestinal lesion has a sensitivity of 80-100% in the diagnosis.¹⁸⁻¹⁹

CT findings in ectopic pancreatic rests are not specific although these lesions can enhance to the same degree as the normal pancreas with IV contrast.³ Newer technologies like capsule endoscopy and single bowel enteroscopy are being used to detect lesions in the small intestine which were previously not accessible without surgery.³ Even though endoscopy recognizes a lesion, superficial biopsies are unable to reveal a submucosal lesion and shows only chronic inflammation.

After other more common causes of abdominal complaints are excluded, symptomatic lesions should undergo surgical resection. However, controversy exists in the management of asymptomatic and incidental lesions.²⁰ Some suggest frequent endoscopy for asymptomatic lesions while others say that there is

no need of increased follow up.³ In those lesions where malignant transformation is seen or malignancy is uncertain, then surgical resection is advocated.²⁰⁻²¹

Conclusion

Though a rare entity, pancreatic heterotopias should be considered in the differential diagnosis of submucosal gastrointestinal lesion. The diagnosis of ectopic pancreas is still challenging in spite of the improvement and development in the diagnostic modalities. Symptomatic relief is provided by surgical resection and surgery is recommended particularly if diagnostic dilemma exists.

Table 1: Characteristic features of cases with heterotopic pancreas

Case	Age	Sex	Symptom	Location	Size	Microscopic type
1	25	M	Pain	Gastric antrum	1cm	Type I
2	28	M	Pain	Gastric antrum	2cm	Type I
3	32	M	Pain	Gastric antrum	1.5cm	Type I
4	35	F	Pain	Gastric antrum	1.5cm	Type I
5	48	F	Pain	Gastric antrum	2cm	Type I

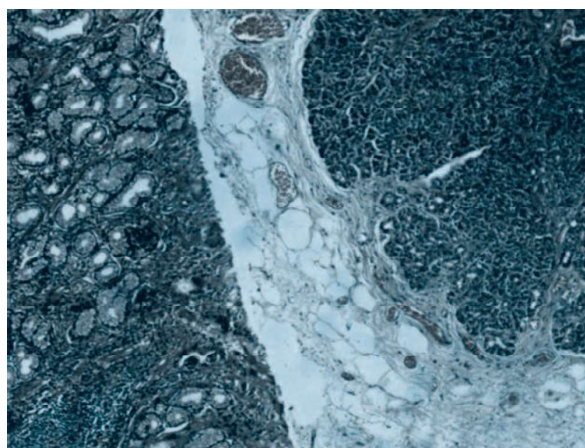


Fig 1. Pancreatic ducts and acini in the pyloric antrum (H & E, 10x).

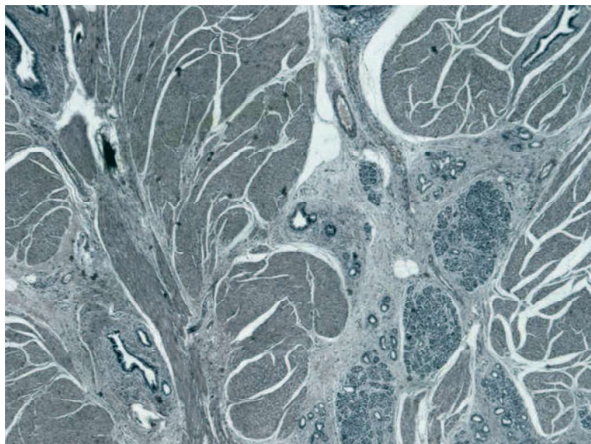


Fig 2. Pancreatic ducts and acini involving the muscularis propria (H & E, 20X).

Conflicts of interest: None declared

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