

# Mucinous cyst exhibiting severe dysplasia in gastric heterotopic pancreas associated with the gastrointestinal stromal tumour

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## Abstract

Heterotopic pancreatic tissue within the stomach is rare and dysplasia within heterotopic pancreatic tissue is very rare. We present the first report of a patient with concurrent occurrence of heterotopic pancreas in the stomach with a gastrointestinal stromal tumour.

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**Key words:** Dysplasia; Gastrointestinal stromal tumour; Heterotopic pancreas; Mucinous cyst; Stomach

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## INTRODUCTION

Heterotopic pancreas is a relatively common lesion found in approximately 1 in 500 abdominal laparotomies<sup>[1]</sup> with approximately 62% found in the stomach<sup>[2]</sup>. Pancreatic heterotopia is defined as pancreatic tissue lacking vascular or anatomical continuity with the normal pancreas<sup>[1]</sup>. Symptoms associated with heterotopic pancreas are rare, but when present are usually associated with a gastric site. Symptoms may be due to mass effect such as pyloric obstruction,

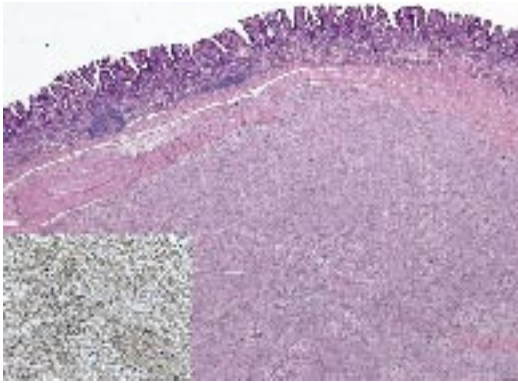
ulceration or bleeding, or due to pancreatic diseases such as pancreatitis, cyst formation or pancreatic neoplasia. Dysplasia and malignancy within pancreatic heterotopia are rare events<sup>[3]</sup>. The synchronous occurrence of gastrointestinal stromal tumour (GIST) and heterotopic pancreas has not, to our knowledge, been previously reported.

## CASE REPORT

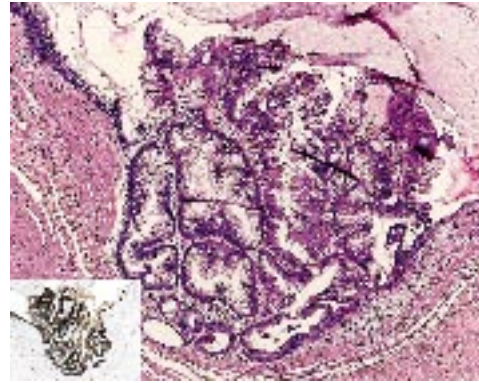
A 71 year-old female was admitted to our hospital for melaena (77 g/L haemoglobin). She underwent endoscopy, and two lesions were visualised in the gastric antrum, one with overlying mucosal ulceration. The lesions were not biopsied as they were scheduled for resection. On the following day, both lesions were resected. Macroscopically the lesions measured 70 mm × 50 mm × 40 mm and 15 mm × 15 mm × 12 mm. The former was a solid tumour with a fleshy tan cut surface, while the latter was a solid and cystic lesion containing a single cyst (10 mm in diameter) filled with mucinous material. The solid lesion with surface ulceration was a gastrointestinal stromal tumour (intermediate risk, uncertain malignant potential)<sup>[4]</sup> composed of fascicles of spindle cells exhibiting mild nuclear pleomorphism and up to 4 mitotic figures per 50 high power fields (Figure 1). Large areas of tumour necrosis were present. Surface ulceration was present, which was the likely source of melaena.

The smaller lesion was composed of pancreatic ducts and acini within the gastric submucosa and focally extended into the muscularis propria. Within this area was a cyst partly lined by mucinous epithelium while in some areas the lining epithelium was denuded. The mucinous epithelium exhibited moderate to severe dysplastic cytoarchitectural features (tufted and micropapillary architecture, along with nuclear enlargement, stratification, coarsely granular chromatin and nucleolar prominence) (Figure 2). Around the cyst there were small pools of extracellular mucin, associated with a chronic inflammatory response suggestive of partial rupture. Adjacent smaller dilated ducts were lined by non-dysplastic columnar mucinous epithelium. No evidence of stromal invasion was present. The cyst lining cells were positive for CK7 and CK20 (Figure 2), the latter was localized in the dysplastic areas. The intracytoplasmic mucin was positive for alcian blue pH 0.5 and periodic acid Schiff.

The gastric mucosa adjacent to the lesions showed active chronic gastritis, *H. pylori* organisms and intestinal metaplasia.



**Figure 1** Gastrointestinal stromal tumour composed of fascicles of spindle cells. The tumour cells are positive for CD117 (inset).



**Figure 2** Severely dysplastic mucinous epithelium lining mucin-filled cyst in heterotopic pancreas. The dysplastic epithelium is positive for CK20 (inset).

## DISCUSSION

Heterotopic pancreas is a relatively common lesion most often present in the gastric antrum seen macroscopically as a round or lobulated white to yellow which can be up to a few centimetres in dimension<sup>[2]</sup>. Diagnosis is difficult and often not made until surgical removal of the lesion. Heterotopic pancreas has been classified into three types by Heinrich: class I is typical pancreatic tissue with acini, ducts and islet cells, class II shows a large number of acini and few ducts, and class III shows numerous ducts with few acini or islet cells<sup>[5]</sup>. Neoplasms arising in heterotopic pancreatic tissue are rare<sup>[6]</sup> and include borderline mucinous cystic tumour<sup>[7]</sup>, adenocarcinoma<sup>[6]</sup>, mucinous cystadenocarcinoma<sup>[8]</sup>, acinar cell carcinoma<sup>[2]</sup>, islet cell tumour<sup>[9]</sup>, or solid and papillary neoplasm<sup>[10]</sup>. Cystic degeneration without malignant change appears to be more common and may mimic mucinous carcinoma from another primary site<sup>[11]</sup>. The case report by Naqvi *et al*<sup>[7]</sup> includes a description of jejunal pancreatic heterotopia with cystically dilated ducts lined by mucinous epithelium showing low grade dysplastic cytoarchitectural features. They diagnosed a borderline mucinous cystic tumour without documenting the presence of ovarian type stroma. Of the reported cases of malignancy few have included reference to dysplastic or pre-malignant change<sup>[8,12]</sup>. These reports note the presence of dysplasia or carcinoma *in-situ* within the heterotopic pancreas adjacent to invasive ductal adenocarcinoma. Given the morphological appearances of the severe dysplasia seen in our patient, it is likely that the changes represent a pre-malignant change akin to that reported in orthotopic pancreas under the rubric “pancreatic intraepithelial neoplasia” (PanIN). Although malignant change within heterotopic pancreas is rare, we recommend that in the presence of dysplastic change within heterotopic pancreas tissue, the entire lesion should be sampled and examined histologically to exclude the presence of invasive malignancy.

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