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Gastric heterotopic pancreas masquerading as a stromal tumor: A case report

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Abstract: **Heterotopic pancreas is a rare condition and its preoperative diagnosis is difficult. It is generally asymptomatic, but it may become clinically evident depending on the size, location, pathological changes and complications similar to those observed in the normal pancreas .Heterotopic pancreas can occur anywhere in the gastrointestinal tract. Most commonly seen in the duodenum, stomach, and jejunum.**
We report a case of a 27 -year-old female patient with epigastric pain due to ectopic pancreatic tissue in the stomach is described. The difficulty in making an accurate diagnosis is highlighted. Distal gastrectomy was performed and a histological diagnosis of heterotopic pancreas was established on microscopic examination. The patient had an uneventful postoperative course and remained symptom free in the follow-up of six months. Areview of the literature on this pathology is hereby presented. Although heterotopic pancreas is rare, it should be considered in the differential diagnosis of pancreatitis and of a submucosal gastric tumour.

Keywords: **ectopic pancreas; stomach; pancreas; histology; heterotopic pancreas; gastrectomy.**

I. INTRODUCTION

Pancreatic heterotopia was first described in 1727 when it was found in an ileal diverticulum[i].It is a rare entity, defined as the presence of extrahepatic tissue without any anatomic or vascular continuity with the pancreas. It may occur at a variety of sites in the gastrointestinal tract having a propensity to affect the stomach and small intestine. Usually, it is a silent anomaly but it may become clinically evident when complicated by inflammation, bleeding, obstruction ,malignant transformation or pseudocyst formation [ii] [iii]. Heterotopic pancreatic tissue is found in persons of all ages and slightly more often in male[iv] . This is a report of a case of non specific epigastric pain resulting from pancreatic heterotopias in the gastric antrum in an adult woman.

II. CASE REPORT

A 27 years – old women was presented to our hospital due to a 4 weeks history of recurrent epigastric pain with no meal relation and no relieving factors .Along with epigastric fullness sensation and intermittent nausea . physical examination , routine blood tests including amylase , plain chest and abdominal x ray were unremarkable . Abdominal ultrasound (figure 1) done showed a defined lesion in relation to the distal stomach with central necrosis and well defined margins , measures approximately 3.9 x 3.1 cm with a tiny echogenic peripheral foci probably calcification, shows minimal vascularity on Doppler imaging in the periphery.; pancreas, liver and spleen were normal. Computed tomography (figure 2) was done and showed a round well defined 3.6 x 3.4 cm hypodense lesion which is probably submucosal. There is no associated lymphadenopathy. The pancreas, adrenals, spleen and both kidneys are within normal limits.

Endoscopic ultrasound showed an intramural (subepithelial) lesion was found in the antrum of the stomach . the lesion was appeared to originate from withi the muscularis properia (layer 4) Tiussue was obtained from this exam through FNA and resulted in (begin gastric epithelial cell and was negative for malignancy) .A decision was made to proceed with surgery . The patient underwent exploratory laparotomy,through a small midline incision , a solid mass around 3 by 3.5 cm located in pre -pyloric area, with a distal limit of 3mm from the pylorus was found.Due to the presumed diagnosis of GIST , the lesion was removed with distal gastrectomy with Billroth II anastomosis was carried out.

The postoperative course was uneventful and she was discharged home 5 days after surgery. She remained free of symptoms and she returned back to her work after 3 weeks.

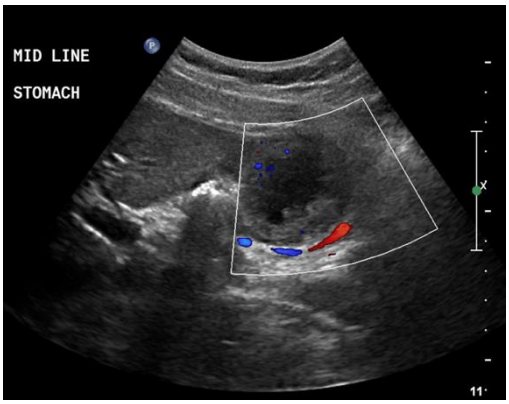

Figure 1 : abdominal US

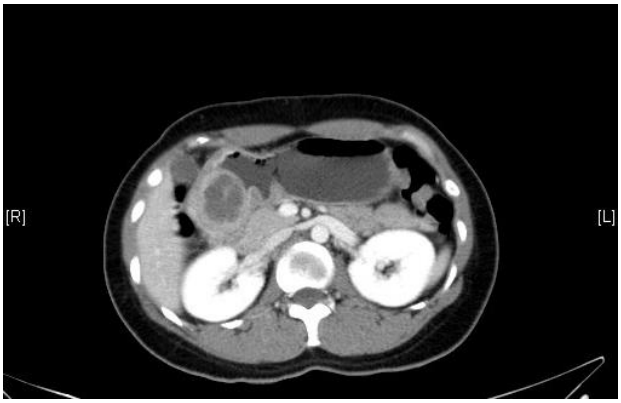

Figure 2 : Abdominal CT scan

Histopathologic examination of the lesion reveal islands of pancreatic acini and ducts in the muscularis propria of the stomach. Several dilated ducts are seen extending through all the layers of muscularis propria. Surrounding the ducts are layers of fibroblastic reaction and scant mixed inflammatory infiltrate. The pancreatic lobules were located mainly in the gastric muscularis propia with normal overlying mucosa Other areas show more extensive fibroinflammatory reaction admixed with histiocytes and few aggregates of giant cells. No evidence of malignancy is seen , resection margins were microscopically negative and the lymph nodes were benign .Immunostains was performed : Cytokeratin 7 : positive in the pancreatic tissue and ducts , CA19-9 positive in the pancreatic tissue and ducts , CD117,CD30 ,chromogranin , ER,PR,CDX2,CK20 all was negative

Concluded histopathological diagnosis heterotopic pancreatic tissue in the gastric antrum with a lobular architecture characteristic of ectopic pancreas.

III. DISCUSSION

The first case of heterotopic pancreas developed in an ileal diverticulum, was reported by Jean Schultz in 1727, but the first histological confirmation was described by Klob in 1859[v]. The incidence in autopsies ranges 0.5%-13.7%, being more common at the age of 30-50 years with a male predominance[vi]. Although its cause is unclear, several theories, including the 'theory of metaplasia,' the 'theory of misplacement' and the latest addition, the 'theory of abnormalities of notch signaling', have been proposed to explain the pathogenesis and occurrence of pancreatic heterotopia [vii viii ix].

Histopathologically, it is not a diagnostic problem when pancreatic acini, ducts, islets of Langerhans and intervening connective tissue are present. The most characteristic gross feature is a central ductal orifice [x]

Specifically in the stomach, the involvement of submucosal layer, muscularis and subserosal layer is 73%, 17% and 10%, respectively[xi] . In this reported case the pancreatic tissue involve muscularis propria . The most common site of this entity is the stomach, accounting for 25-38% of cases, followed by the duodenum (17-36%) and jejunum (15-22%). Reported rare cases of heterotopic pancreas were found in the esophagus, common bile duct, gallbladder, mesentery, spleen, mediastinum or fallopian tubes . Heterotopic pancreas is usually an isolated lesion, although multiple lesions have also been observed [xii]

The pancreatic ectopic tissue is usually silent but can also undergo complications that occur in normal pancreatic tissue such as acute or chronic pancreatitis, abscess and pseudocyst formation [xiii], Malignant transformation may rarely occur[xiv]. Symptoms depending upon the location, such as gastric outlet obstruction in a pre-pyloric or obstructive jaundice in a bile duct focus and are also related to the size of the lesion. Symptoms frequently seen in Lesions greater than 1.5 cm in diameter[xv]. Pain is one of the most common symptoms. The possible explanation is that the pain is due to the secretion of hormones and enzymes, being responsible for inflammation or chemical irritation of the involved tissues[xvi]. In certain cases, ectopic pancreases manifest as conditions that require emergency treatment, such as gastrointestinal bleeding, gastric outlet obstruction or perforation These cases require further management involving surgery or endoscopical intervention. [xvii]

Although modern medicine offers a variety of diagnostic methods, the diagnosis of heterotopic pancreas remains a significant challenge; this has been demonstrated by the frequent inability to differentiate heterotopic pancreas from neoplastic lesions warranting surgical excision. Imaging techniques such as EUS and CT are frequently used for

gastrointestinal submucosal tumor diagnosis and may be of use in the diagnosis of gastric heterotopic pancreas but are not sufficiently specific for differential diagnosis. Endoscopically, heterotopic pancreas in the stomach wall has been described as an elevated delomorphic submucosal tumor that presents with a normal overlying mucosa, with characteristic central umbilication [xviii] . However, in more than half of recorded cases, the endoscopic view is not that specific, for example, the central dimpling is missing, and the tumor may therefore easily be misinterpreted as another submucosal tumor [xix] . On EUS, Heterotopic pancreas is typically hypoechoic and heterogeneous, with indistinct margins and usually arises from the second, third and/or fourth layers of the gastrointestinal tract, or from a combination of the three [xx] Meanwhile, GISTs are typically hypoechoic, homogeneous lesions with well-defined margins. However, these tumors can also occasionally present with ulcerations and irregular margins. The majority of GISTs originate from within the muscularis propria. Small lesions may originate from the muscularis mucosa [xxi]. Although the imaging features of an ectopic pancreas, including a larger longest:shortest diameter ratio, a mural growth pattern, an antral location, third (submucosal) layer disruption, intermediate echogenicity and irregular margins, can occasionally aid in distinguishing heterotopic pancreas from other submucosal gastrointestinal tumors, none of these findings, either individually or collectively, are characteristic of this entity, and no long-term studies have supported this description. Therefore, a surgical resection is inevitably required to confirm the diagnosis in the majority of circumstances [xxii xxiii]

The definitive diagnosis can be reached by the histopathological examination of the tissue. EUS-FNA has emerged as an important diagnostic technique for submucosal tumors [xxiv]. However, as this method exhibits limited diagnostic accuracy due to the small amount of tissue that can be collected with a chance of non-diagnostic sample, an improved method, such as ESD-based intervention or surgery, is required for tissue sampling [xxv].

Opinions concerning management in case of ectopic pancreas are inconclusive. Some authors believe that surgery is required, while others believe that the lesion should be monitored, especially if asymptomatic, with surgical intervention only in case of complicated cases and neoplastic transformation [xxvi xxvii]. At present, selecting the optimal treatment is a challenge even for experienced clinicians and many authors are considering surgical intervention is recommended in patients manifesting clinical symptoms, as to exclude neoplastic lesions. Three methods of treatment intervention currently exist: Classic laparotomy with wedge resection, laparoscopic resection and endoscopic submucosal dissection (ESD) [xxviii xxix xxx].

The diagnosis may be sometimes challenging intraoperatively due to the gross similarity of pancreatic heterotopia with gastrointestinal stromal tumor (GIST), gastrointestinal autonomic nerve tumor (GANT), carcinoid, lymphoma or even gastric carcinoma. If in doubt, frozen section is very helpful to establish the diagnosis intraoperatively and to avoid unnecessary extensive operations.

In our opinion, a precise pre-operative diagnosis through imaging and pathology, which may provide useful information, should be performed prior to deciding on the therapeutic strategy. EUS-FNA is useful for making an accurate histological diagnosis of the lesion, although its value requires further assessment and future improvements. In addition, an ESD-based method can overcome the flaw of no adequate tissue samples being obtained from conventional biopsy and EUS-FNA, and can deeply improve the accuracy of diagnosing this disorder prior to management .

IV. CONCLUSION

Heterotopic pancreas should always be considered when diagnosing extramucosal gastric masses. Precise pre-operative diagnostics may avoid misleading and unnecessarily excsions. Endoscopic submucosal dissection-based management in ectopic pancreases deserves further consideration and investigation in the future.

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