Case Report

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Ectopic Pancreatic Islets in Splenopancreatic Ligament: A Case Report

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ABSTRACT

Heterotopic or ectopic pancreas is pancreatic tissue present at an abnormal location which is not connected to the main pancreas. They are predominantly located in the gastrointestinal tract, with stomach being the most common site. Mostly asymptomatic, they are usually detected incidentally during surgeries for other conditions. Preoperative diagnosis is very difficult and the definitive diagnosis is histopathological examination of the suspected lesion. Surgical resection is curative and it should be done in asymptomatic cases for diagnostic confirmation and prevention of complications. Here, we present a case of ectopic pancreas in a 59-year-old female in the splenopancreatic ligament which was incidentally identified during histopathological examination of specimen excised for serous cystadenoma of pancreas.

Keywords: *Ectopic, Serous Cystadenoma, Incidental, Langerhans, Splenopancreatic Ligament*

Introduction

Heterotopic or ectopic pancreas (EP) is the presence of pancreatic tissue at any abnormal location which has no anatomic, vascular or neural continuity with the main pancreas [1]. It is believed to be a developmental anomaly occurring during foregut rotation where small amounts of pancreatic tissue gets detached and develop at aberrant locations [2]. They are generally incidental findings with the most common sites being stomach, duodenum, jejunum [3,4] and rarely umbilicus [2], Meckel's diverticulum [5] and mediastinum [6]. The frequency of occurrence is 1 in 500 laparotomies [7]. Ectopic pancreas usually presents during 5th or 6th decades of life and has a male preponderance. Most patients with ectopic pancreas present without any symptoms and the diagnosis is often made incidentally during surgical exploration for other causes. Few imaging modalities have been suggested to aid the preoperative diagnosis of heterotopic pancreas but histological examination of the suspected lesion still remains the gold standard for definitive diagnosis [8]. Grossly, it may present as a small firm, pale, nodular mass. Microscopically, there are lobules of morphologically unremarkable pancreatic ducts, acini and frequently, but not always, islets of Langerhans cells. Since it contains normal pancreatic tissue, complications like acute pancreatitis or rarely neoplasms like mucinous cystadenoma or cystadenocarcinoma may arise [9]. Complete surgical resection is the treatment of choice in these cases. Herein, we present a case of ectopic pancreatic islets diagnosed incidentally in the splenopancreatic ligament in a case of serous cystadenoma of pancreas.

Case Presentation

A 59-year-old lady, known diabetic, was admitted with the complaints of fever with chills and associated generalized weakness for 2 days. Investigations following admission revealed neutrophilic leukocytosis in complete blood count (total WBC count=17310/µL) and E. Coli in urine culture. USG whole abdomen done outside showed a hypoechoic pancreatic mass. Biopsy taken from the mass was predominantly necrotic without any viable pancreatic tissue. But endoscopic USG guided FNAC showed occasional clusters of suspicious atypical cells.

Further workup included CT whole abdomen which showed a mass in the tail of pancreas with a maximum dimension of 6 cm. This was followed by MRCP revealing a well-defined multiloculated cystic lesion in the tail of pancreas with a maximum size of 7.6 cm with multiple thin internal septations. Few of the septations showed specks of calcification. PET CT showed the lesion to be mildly FGD avid (SUV max 3.4). (FIG 1)

She underwent laparotomy. During surgery, the lesion along with the tail and part of body of the pancreas, splenopancreatic ligament and spleen were removed and sent for histopathological examination.

Histopathological Examination

The histopathological specimen was that of distal subtotal pancreatectomy and splenectomy which all together measured 14.5 x 8.5 x 4 cm. The spleen measured 9 x 7.5 x 2.5 cm and weighed 67.5 grams. The portion of pancreas measured 9.5 x 8.3 x 4.4 cm. There was a pale grey to white

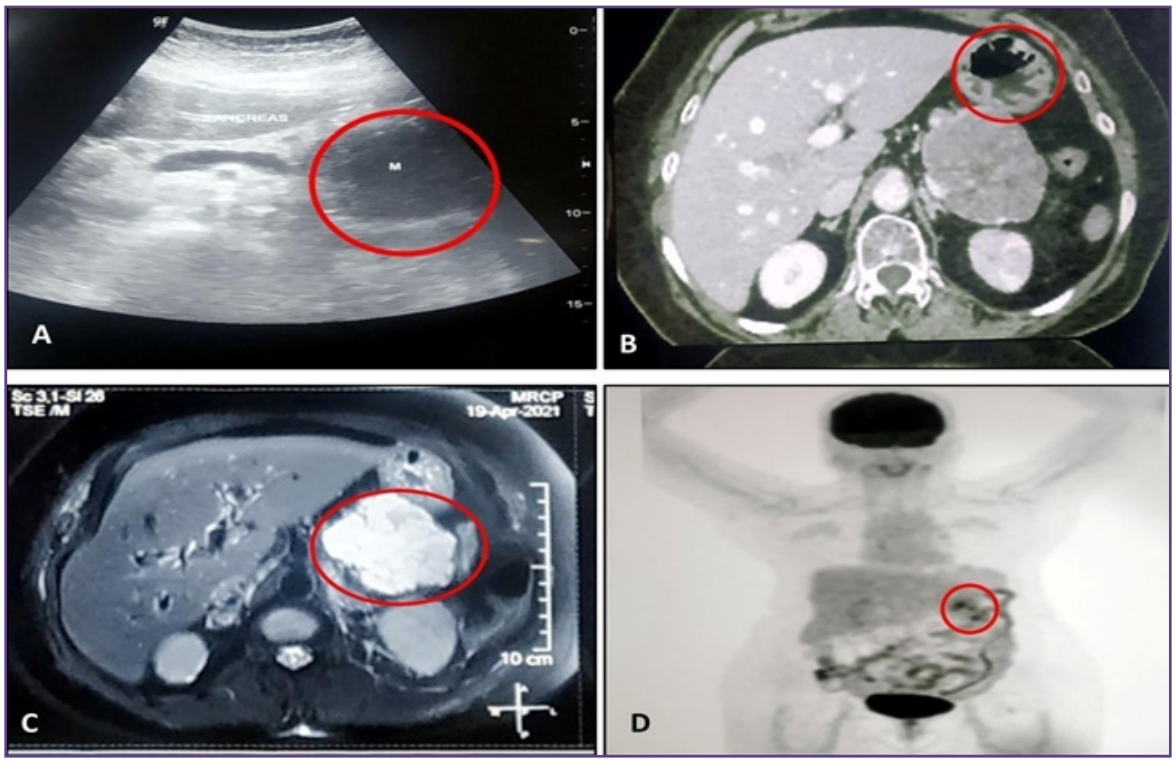

Fig. 1: Imaging findings of the patient (red circle indicates the lesion) A: USG B: Plain CT whole abdomen C: MRCP D: PET CT

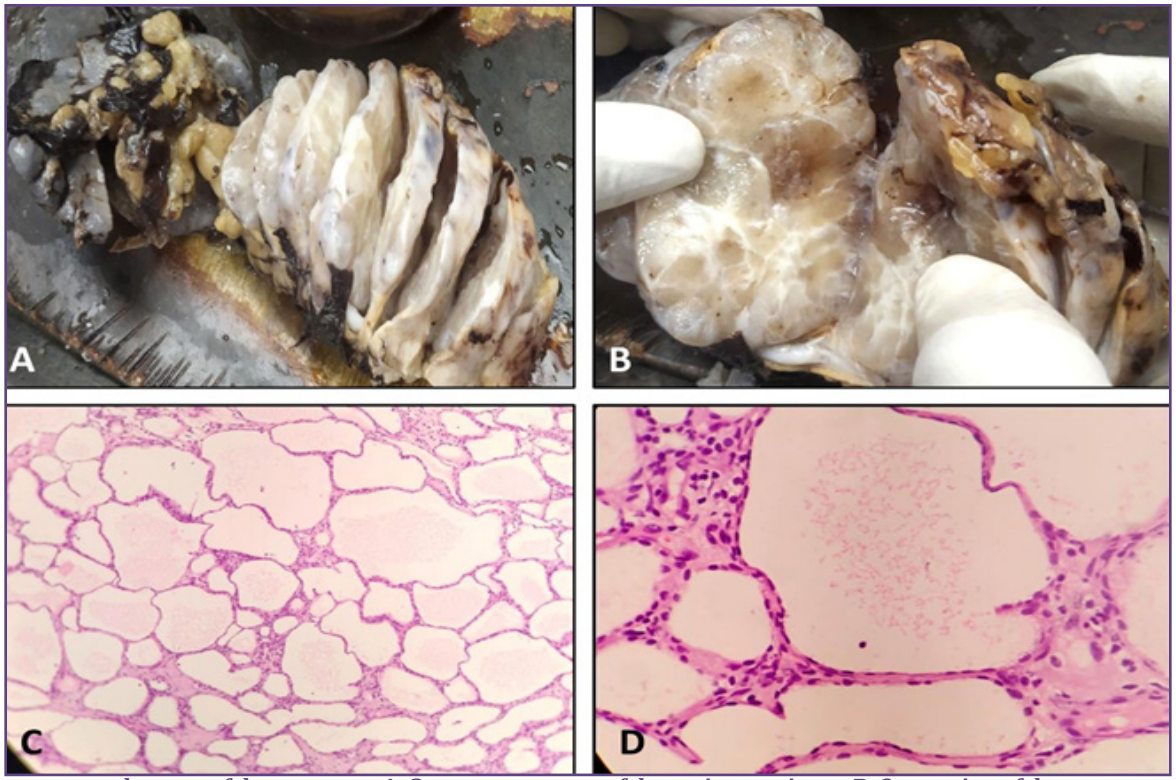

Fig. 2: Serous cystadenoma of the pancreas. A: Gross appearance of the entire specimen. B: Cut section of the pancreas showing multicystic appearance. C: Multiloculated pancreatic cyst lined by flattened cuboidal epithelium (100x magnification) and D: Appearance of the cyst at 400x magnification.

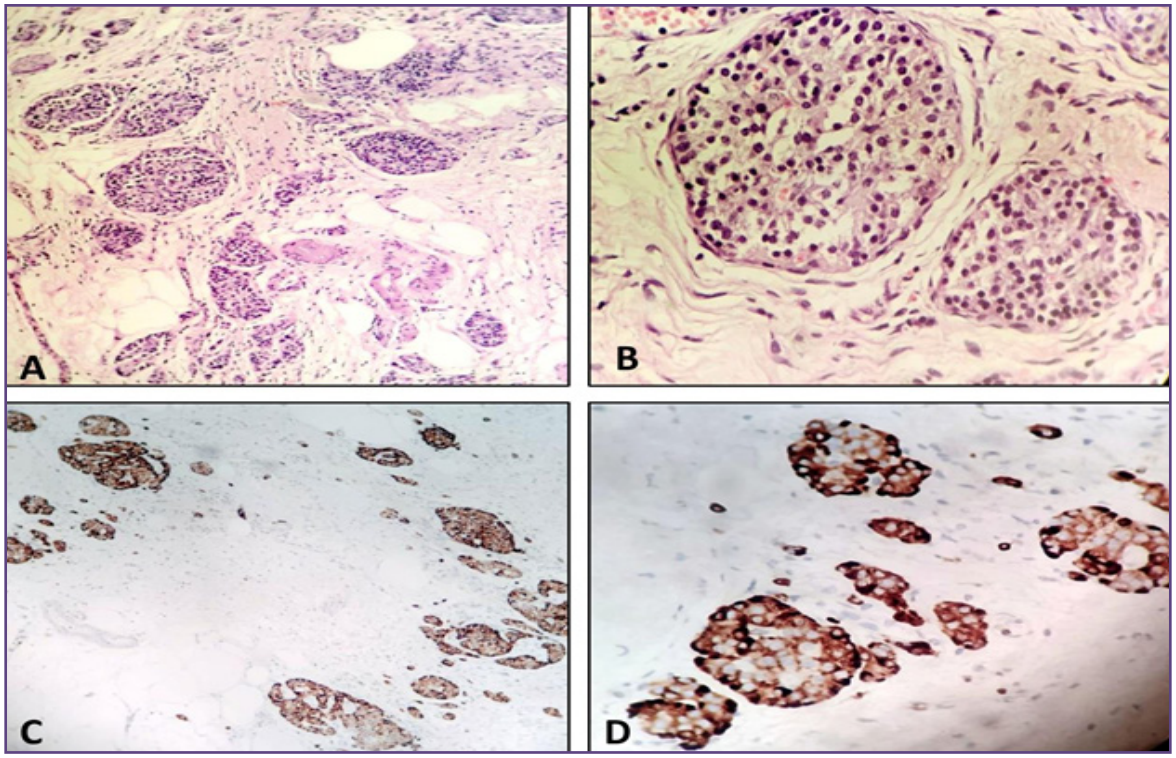

Fig. 3: Ectopic pancreatic islets in splenopancreatic ligament. A: 100x and B: 400x magnification. C: Positivity for Synaptophysin. D: Positivity for chromogranin.

well circumscribed, lobulated, firm mass in the pancreatic body and tail measuring 8 x 7 x 4.4 cm which was 2 cm from pancreatic neck cut margin. The cut section of the mass was solid to cystic with hemorrhagic areas (FIG 2-A, B). Multiple sections from the lesion, normal pancreas, and spleen were taken. Lymph nodes from splenic hilum, splenopancreatic ligament and peripancreatic fat were taken and processed for microscopy

Microscopical Examination

Microscopic examination of the lesion showed multicystic appearance with cysts lined by cuboidal to flattened epithelium and areas of dystrophic calcification were noted. There was no dysplasia or malignancy and the findings were consistent with serous cystadenoma of pancreas (FIG 2-C, D). Lymph nodes showed features of reactive hyperplasia.

One of the nodular areas which were sampled as a lymph node showed groups of pancreatic islet cells without accompanying ducts. The nodule measured 0.8 x 0.6 x 0.4 cm and was located in the splenopancreatic ligament. The presence of islet cells was confirmed by immunohistochemistry which were positive for synaptophysin and chromogranin. There was no atypia or malignancy in the ectopic tissue. (FIG-3)

Discussion

Ectopic pancreas is a rare occurrence with various theories being proposed for its origin. Of them, misplacement or poor placement theory proposed by Nakama et al [2] is the most widely accepted. They postulated that portions of primitive pancreatic tissue gets separated during foregut rotation and fusion of the pancreatic buds, which then gets relocated anywhere in the gastrointestinal tract.

The exact incidence of heterotopic pancreas is difficult to ascertain as most patients are asymptomatic. Literature reports a prevalence between 0.5 and 15% in autopsy findings and 0.2% in laparotomies [7]. The incidence can increase to 0.9% in gastrectomies. In a large study done from a single high volume medical centre in China, it was reported that around 31% of all ectopic pancreatic (EP) tissue is detected intraoperatively during procedures for other conditions [10]. In our case, ectopic pancreatic islets were detected incidentally during surgery for serous cystadenoma of pancreas.

Due to the close proximity of primordial pancreatic buds and the foregut, ectopic pancreatic tissue commonly occurs in the upper gastrointestinal tract. The most common site is stomach (24 to 38% of cases), followed by duodenum (9-36%), jejunum (0.5- 27%), ileum (3-6%), and Meckel's

diverticulum (2-6.5%) [3-6] .5-16% of cases occur in the peripancreatic region.

In our case, ectopic pancreatic tissue was found in the splenopancreatic ligament. Gupta and Vasishta in 2008 [11] and Panda et al in 2015 [12] reported ectopic pancreatic tissue in splenic hilum. In patients with ectopic pancreas in gallbladder, bile ducts, splenic hilum, or liver, a higher incidence was observed in middle aged females [12]. In both of the above-mentioned reports, the patients were female- the former being younger with 21 years, the latter being middle aged with 40 years. In our case the female was aged 59 years.

Most of the patients are asymptomatic. When they do become symptomatic, the most frequent presentation is abdominal pain due to inflammation resulting from a release of pancreatic enzymes. Complications are similar to those of the normal pancreas and include pancreatitis, bleeding, ulceration of the overlying gastric mucosa, pseudocyst formation. Malignant transformation of HP is rare (0.7–1.8% of cases) [9,13] Our patient had no symptomatic finding related to the ectopic pancreas.

Usually, the ectopic tissue is small, but it can reach up to 3 cm in dimension [14]. Clinically significant lesions are usually larger than 1.5 cm. Grossly, they present as a firm white nodule. Microscopically, they can be composed of pancreatic ducts, acini and islets of Langerhans. Immunohistochemistry shows CK7, CK19 positivity in ducts and acini and synaptophysin chromogranin positivity in the Langerhans cells similar to normal pancreas [1]

Heterotopic pancreas is classified based on the modification of Heinrich criteria [15]:

- Type 1: presence of acini, ducts, and islet-like pancreatic gland.
- Type 2: canalicular variant with pancreatic ducts.
- Type 3: exocrine pancreas with acinar tissue.
- Type 4: endocrine pancreas with cellular islets.

Initial classification of Heinrich had only the first 3 classes with the fourth class being introduced by Gaspar in 1973. Of all the classes, type 1 is the most common. Our case belonged to the type 4 category

It is difficult to preoperatively diagnose EP. A few imaging modalities like endoscopy +/- ultrasound, CT, magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) may aid in preoperative diagnosis [8]. However, histological examination of the suspected lesion is the method for definitive diagnosis.

Local excision is the appropriate management for EP [14]. Surgical excision can provide tissue for histological diagnosis and can serve to rule out malignancy. Additionally, surgical excision may prevent future complications like pancreatitis, pseudocyst formation or malignant transformation. Hence, identification of the lesion and its removal remains of paramount importance even in asymptomatic patients.

Conclusion

It is concluded that peripancreatic fat and splenopancreatic ligament can be a rare location of ectopic pancreas. The lesion can easily be missed on radiology or may be interpreted as a reactive lymph node or neoplastic foci. Hence, surgical removal is recommended when found incidentally. This will help in histopathological confirmation of the lesion and also prevent future complications.

Patient consent: Though this report does not reveal any details of the patient, we separately collected informed patient's consent for reporting this case.

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Declarations of Competing Interest

The authors have no conflicts of interest to declare.

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