

A rare case of ectopic pancreas in the ampulla of Vater presented with obstructive jaundice



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A very rare case of ectopic pancreas in the ampulla of Vater presented with obstructive jaundice

A rare case of ectopic pancreas in the ampulla of Vater presented with obstructive jaundice. Ectopic pancreas is a common congenital disorder which is referred to as pancreatic rest. The incidence in autopsy series varies from 1 to 2% (range 0.55 to 13%). Most common site of ectopic pancreatic tissue is the stomach, although it can be found anywhere in the foregut and the proximal midgut.

Ectopic pancreatic tissue in the ampulla of Vater is a very rare condition. Searching in the literature, using the terms "ectopic pancreas" and "Ampulla of Vater", we found only 10 records.

The authors report on a rare case of a 69 years old female with ectopic pancreas in the ampulla of Vater, presented with painless obstructive jaundice, and the diagnostic and therapeutic strategy that followed.

KEY WORDS: Ampulla of Vater, Ectopic pancreas, Obstructive jaundice

Introduction

The presence of pancreatic tissue lacking anatomic and vascular continuity with the main gland is called ectopic or heterotopic pancreas and can be located in various anatomical sites related with the foregut and proximal midgut. This lesion usually presents no symptoms¹⁻³. Most common sites are the stomach, the duodenum and the jejunum which account for about 75% of all ectopic pancreatic tissue. Other less common sites are the ileus, the Meckel's diverticulum, the gallbladder etc². This entity is identified usually in autopsies and the frequency ranges from 0.5% to 13% between several series. The rate of recognition at the time of laparotomy is 0.2%. Ectopic pancreas may cause problems in the differential diagnosis during endoscopies. (1, 2, 3) Ectopic pancreas in the ampulla of Vater presenting with obstructive jaundice is a very rare entity. We searched the literature using as key words: "ectopic pancreas" and "ampul-

la of Vater" and we found only 10 records. Heterotopic pancreas may cause intra-abdominal bleeding as leading symptom. The treatment of choice proposed by the authors was resection of the ectopic pancreatic tissue with the distal common bile duct with the ampulla and anastomosis of the common bile duct with the duodenum.

Case Report

A 69 years old female proceeded in the Emergency Department of our Hospital complaining for painless jaundice and weight loss. The patient's history was clear. Blood analyses as well as imaging exams were performed. The blood analysis was normal, and only the total and direct serum bilirubin were increased. Cholelithiasis with a mild distension of the common bile duct was the only finding in the U/S. The CT scan revealed a mass or an oedema with not definite characteristics, so without defining the origin of it. Although cholelithiasis was present in the U/S, CT imaging failed to reveal bile duct lithiasis.

ERCP was performed for diagnostic and therapeutic purposes because serum bilirubin levels was significantly increased. ERCP revealed a stenosis in the ampulla of

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Vater probably due an external pressure. The endoscopist failed to perform endoscopic sphincterotomy or to place a stent for drainage of the common bile duct, which was dilated. (Fig. 1). The pathologic examination of the biopsies received during the ERCP revealed normal epithelium. MRCP was performed but failed to facilitate the differential diagnosis (Fig. 2). Pancreatic cancer or cholangiocarcinoma was considered as the most possible diagnoses, and the patient informed that may receive a Whipple procedure. Laparotomy was performed and a mass was identified inside the duodenum. The anterior wall of the duodenum was opened and biopsies were done from an intramural mass in the ampulla of Vater, which was at this time considered to be a gastrointesti-

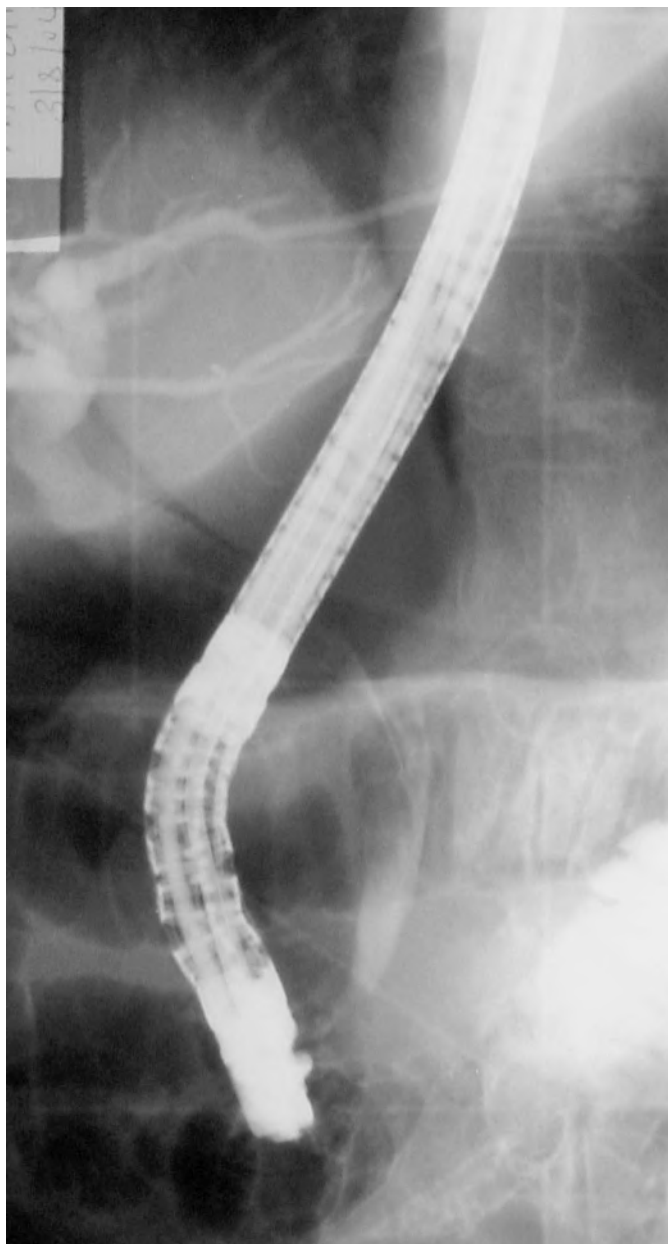


Fig. 1: ERCP demonstrating the stenotic lesion of the distal part of the common bile duct without choledocholithiasis.

nal stromal tumour of the duodenum. The cold biopsy revealed instead ectopic pancreatic tissue. The ectopic pancreas was located near the ampulla obstructing the common bile duct by pressuring its distal part. The mass was then locally resected, with preservation of the ampulla and the distal common bile duct.

The location of the ectopic pancreas facilitated the local excision with preservation of the ampulla of Vater. After the removal of the heterotopic pancreatic tissue the duodenal wall was sutured with absorbable sutures (vicryl 3.0) in two layers.

Bilirubin levels decreased steadily and returned to normal the 7th postoperative day. The postoperative course was uncomplicated and the patient returned home the 10th day. The histological analysis of the specimen confirmed the initial diagnosis set by the cold biopsy (Fig. 3).

A year after the operation the patient is free of symptoms.

Discussion

The pancreas develops from the two primordial diverticula of the duodenum after the 4th gestational week. During the organogenesis of pancreas a number of embryological abnormalities mistakes may occur. Congenital anomalies of the pancreas include ectopic or heterotopic pancreas³. The exact mechanism for this congenital abnormality is not very clear. The embryologists believe that multiple different mechanisms may account for this anomaly. Most accepted hypothesis is that of Horgan and Whartin who suggest that adhesion of portions of the dorsal and ventral pancreatic primordia to the neighbouring structures during elongation and rotation of the gut may be responsible for ectopic pancreas development². Pancreatic buds rising from the embryonic development can attach the gut wall and lead to heterotopic pancreas development. This close relation to embryonic buds of pancreas with foregut is obviously associated with the fact that more than 90% of the cases of ectopic pancreas are located in the upper gastrointestinal tract.

Most commonly ectopic pancreas is located in gastric antrum, gallbladder, liver, spleen, omentum etc.

We searched Medline database for cases of periampullary ectopic pancreas and only 10 records were found. Ectopic pancreas in the ampulla of Vater is an extremely rare entity.

The most common symptoms associated to ectopic pancreas are epigastric pain, dyspepsia, bleeding, acute pancreatitis, pyloric obstruction, cyst formation and inflammation of the adjacent tissues²⁻⁶.

In our case the leading symptom was painless obstructive jaundice. The clinically silent rising of the disease in combination with dysphoria, weakness and the weight loss suggest the possibility of a perimampullary malignancy.

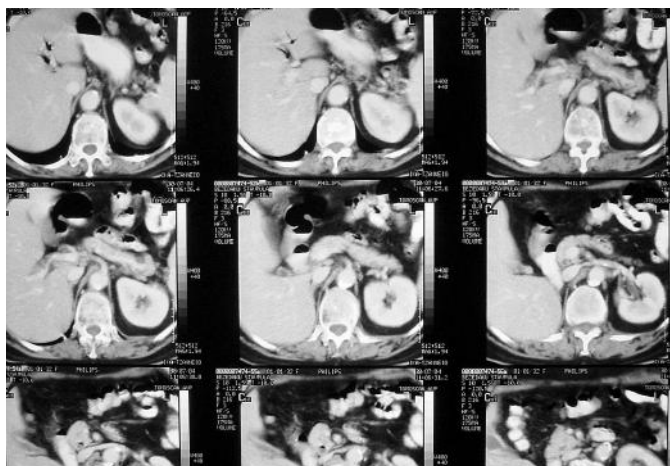


Fig. 2: MRI failed to demonstrate choledocholithiasis although cholelithiasis was present.

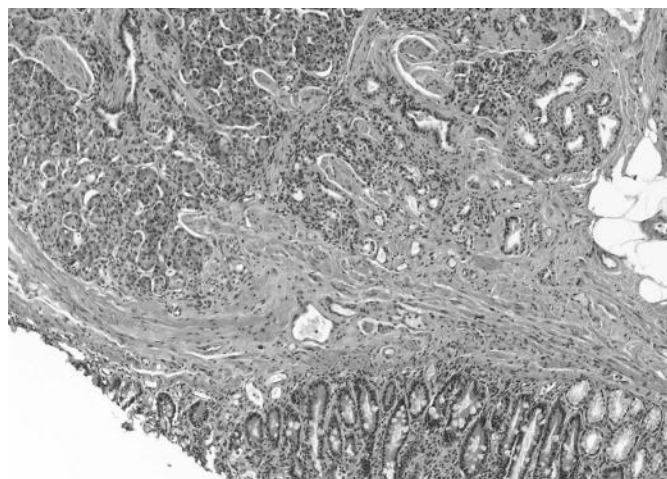


Fig. 3: The histological examination of the removed specimen revealed ectopic pancreas in hematoxylin-eosin stain.

Although most of the authors support the diagnostic and therapeutic role of endoscopy in our case gastroscopy failed to reveal the tumour suggesting an intramural tumour ⁷.

The appropriate management of ectopic pancreas is still unclear and controversial. In most of the cases ectopic pancreas is an asymptomatic incidental finding and does not require necessarily excision unless the diagnosis is obscure. Surgical treatment should be reserved for complicated cases with clinical symptom or for the differential diagnosis of the tumour from other lesions as leiomyoma or carcinoid. In such cases the most common procedure is the resection of the tumour with the ampulla anastomosing the common bile duct with the duodenum ⁴⁻⁸.

In our case the tumour was resected with preservation of the ampulla of Vater in order to avoid the anastomosis between common bile duct and duodenum which increases morbidity. The cold biopsy revealed the ectopic pancreas and no further excision was necessary due to benign characteristics of the lesion.

Riassunto

Il Pancreas Ectopico è una nota malformazione congenita di sviluppo al quale si fa riferimento come se fosse un pancreas residuo. L'incidenza nelle serie di casistiche autoptiche varia da 1-2% (con range tra 0.55 e 13%). La sede più frequente di localizzazione del tessuto pancreatico ectopico è lo stomaco ed il tratto digestivo superiore, ma lo si può anche nel tratto medio dell'intestino e un po' ovunque.

Una condizione veramente rara è la localizzazione di tessuto pancreatico ectopico nel contesto dell'ampolla di Vater. Cercando nella letteratura scientifica internaziona-

le, con l'uso contemporaneo delle chiavi di ricerca "pancreas ectopico" e "ampolla di Vater", si trovano solamente 11 riferimenti.

Presentiamo dunque il raro caso di una paziente 69enne con presenza di tessuto pancreatico ectopico nel contesto dell'ampolla di Vater, pervenuta alla nostra osservazione per un quadro clinico di ittero ostruttivo indolore. Si riferisce sulla strategia diagnostica adottata e quella terapeutica relativa.

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