

Diagnostic Challenge in Heterotopic Pancreas in the Ampulla of Vater with Obstructive Jaundice – A Case Report and Literature Review

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Rezumat

Provocări în diagnosticul unui caz de pancreas ectopic la nivelul ampulei Vater cu icter obstructiv - prezentare de caz și revizuire a literaturii

Pancreasul heterotopic este o anomalie congenitală rară. Cele mai comune locații sunt stomacul, duodenul, jejunul și mult mai rar ampula Vater, esofagul, ileonul, diverticulul Meckel, tractul biliar, mezenterul și splina. Prezentăm cazul unei paciente de 49 ani cu icter mecanic prin pancreas ectopic la nivelul ampulei Vater, inițial interpretată ca tumoră malignă. S-a practicat duo-

denopancreatectomie cefalică tip Whipple cu evoluție favorabilă, concentrația serică a bilirubinei normalizându-se după prima săptămână postoperator.

Cuvinte cheie: pancreas heterotopic, icter mecanic, ampula lui Vater

Abstract

Heterotopic pancreas is a rare congenital abnormality. The most common location is the stomach, duodenum and proximal jejunum. Rare locations are represented by the ampulla of Vater, esophagus, ileum, Meckel diverticulum, biliary tract, mesentery and spleen. We present the case of a 49 year old patient investigated for obstructive jaundice and diagnosed with an ampullar heterotopy of pancreas parenchyma, initially considered to be a malignant tumor. A Whipple pancreatoduodenectomy was performed with good postoperative evolution, the serum levels of bilirubin being normal after the first postoperative week.

Key words: heterotopic pancreas, obstructive jaundice, ampulla of Vater

Introduction

Heterotopic pancreas is a congenital anomaly in which the accessory pancreatic tissue is separated from an anatomic point of view from the original organ, without a vascular or ductal continuity (1,2). The first discovered case was presented by Jean Schultz in 1727, the structure being found in a ileum diverticulum during autopsy on a newborn (1,3). Since then, only 22 new cases have been reported so far with ampullar localization (1). The most common locations for the accessory pancreas are the stomach, the duodenum and the proximal jejunum (1,2) while rare locations are represented by ampula of Vater, oesophagus, ileum, Meckel diverticulum, biliary tract, mesentery, lung, mediastinum and even omentum. The aim of the current paper is to present the case of a 49 year old female initially investigated for jaundice in whom the initial suspicion was the one of a pancreatic head adenocarcinoma. However, after resection, the histopathological studies demonstrated the presence of an ectopic pancreatic tissue at the level of the Vater ampulla. Informed consent of the patient was obtained before publishing the paper.

Case Report

The 49 year old patient was initially investigated for jaundice and epigastric pain. The abdominal ultrasound showed a dilated gallbladder containing sludge in association with dilated common bile duct. Biochemical studies demonstrated the presence of a severe cholestatic syndrome – with a total bilirubin of 11 mg/dl and direct bilirubin of 7 mg/dl; however, the serum levels of CA 19-9 and carcinoembryonic antigen – CEA – were normal. The radiography demonstrated a difficult passage of barium at the second segment of the duodenum, while at upper digestive endoscopy the duodenal mucosa was normal; meanwhile, at the level of the Vater ampula, a hyperaemic area was observed, without any other pathological aspects. In this respect, multiple biopsies were retrieved, but with inconclusive results. The patient was further submitted to an abdomino-pelvic computed tomography (CT) and to an upper abdominal magnetic resonance imaging (MRI) revealing an area with low uptake by the head of the pancreas of 5/3.7 cm, in close proximity to the second and third segment of the duodenum highly suggestive for a primary

pancreatic tumor, without vascular invasion; the rest of the pancreatic parenchyma was normal; meanwhile no distant lesions were found excepting certain infracentimetric adenopathies (Fig. 1).

The investigation data predicted an ampullary tumor with associated jaundice; therefore a Whipple procedure was performed with an uneventful postoperative evolution, the patient being discharged in a week; in the day of discharge the cholestatic syndrome disappeared, the serum values of total bilirubin being of 0.9 mg/dl with a direct bilirubin of 0.5 mg/dl. The histopathological studies demonstrated the absence of any malignant cells, the final diagnosis being the one of Fuentes type I complete heterotopic pancreas in the ampulla of Vater (pancreatic acinar cells, ducts and Langerhans islands). The parenchymal ampullar heterotopy was an atrophic one, with a tube like pattern of growth, interlobular fibrosis and vascular congestion, without malignant elements (Fig. 2 A, B).

Discussions

Heterotopic pancreas is localized in the



Figure 1. Abdominal MRI demonstrating the presence of an area with low uptake at the level of the pancreatic head measuring 5/3.7 cm, in close proximity to the second and third segment of the duodenum, without vascular invasion

stomach, duodenum and jejunum in 75% of cases and, less frequently within the gallbladder, biliary tract, Meckel diverticulum and uterine tube (4). Heterotopic pancreas in the ampulla of Vater represents a rare finding, only 22 cases being reported so far in literature; moreover, only 19 of these cases also associated biliary tract dilatation (5-11). Other associated

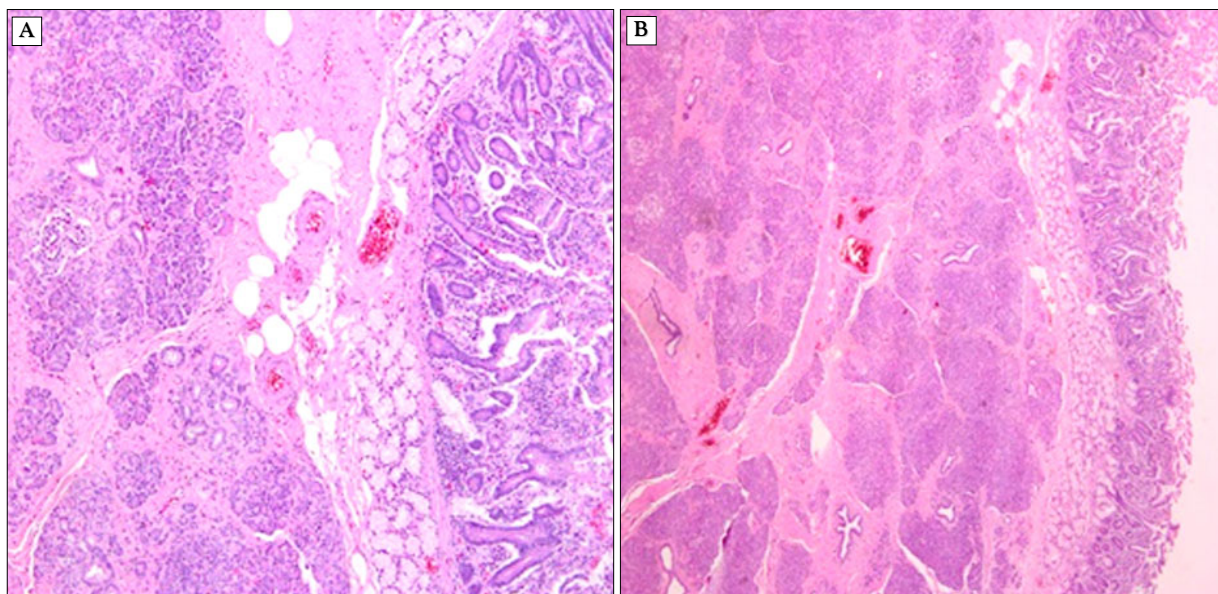


Figure 2. (A, B) Atrophic parenchymal ampullar heterotopy revealing tube like pattern of growth, interlobular fibrosis and vascular congestion, without malignant elements – hematoxylin eosin staining (*10).

conditions are represented by epigastric pain, dyspepsia or bleeding (12).

The tumoral aspect of the papilla makes the diagnostic very difficult and, in most cases, the real diagnosis being established after surgery, based on the histopathological studies. Upper digestive endoscopy can reveal the presence of an ampullary lesion but with a normal aspect of the mucosa; therefore in such cases retrieving a biopsy from this level might not give any significant information (13). In this respect, in certain cases association of endoscopic ultrasound might provide accurate information in order to orientate the possible diagnostic. Therefore, in our case, the initial diagnostic suspicion was the one of a classical pancreatic head tumor associating jaundice and therefore, the therapeutic plan was built accordingly. This therapeutic strategy as well as the diagnostic suspicion seems to be perfectly justified if we take into account the difference in regard to the incidence of the two pathologies: primary pancreatic head adenocarcinoma versus heterotopic pancreas located at the level of the ampulla of Vater. However, we should not omit the fact that association of other investigations such as endoscopic ultrasound might orientate the diagnostic. Therefore, in such cases, a classical endoscopy might show only a central umbilication at the level of the ampulla, while an endoscopic biopsy could not bring any novelty due to the submucosal disposition of the lesion. Meanwhile, adding other imagistic studies such as computed tomography, magnetic resonance imaging or retrograde colangiopancreatography might not increase the accuracy of diagnostic. Association of endoscopic ultrasound seems to bring new information regarding the dimensions of the lesion, the layer of origin, the relationship with the muscularis propria or the coexistence of any pathological lymph nodes while endoscopic ultrasound guided fine needle biopsy might clarify the diagnostic (14-16). However, cases submitted to this kind of therapeutic approach are rather scarce due to the rarity of this suspicion of diagnostic (17).

As for the anatomic-pathological classifica-

tion of this entity, four different types of lesions have been identified so far; type 1 lesions consist of the presence of heterotopic tissues containing all normal pancreatic tissue types – ducts, acinar and insular cells, type 2 lesions – containing only ducts, type 3 lesions – containing only acinar cells and type 4 lesions respectively - containing only insular cells. Malignant tumors arising from this tissue are similar to the ones which can be found in the normally disposed pancreatic parenchyma and consist of ductal adenocarcinomas, cystic adenocarcinomas or acinar carcinomas (14).

Even though, the definitive diagnosis of heterotopic pancreas relies on histopathological examination of the resected tissue. Frozen section analysis during surgery can help guide the surgical approach. Misdiagnosis as a malignancy can lead to unnecessary extensive surgical resection, such as pancreaticoduodenectomy (Whipple procedure), with associated morbidity and mortality. Even though, up to 80% of cases are submitted to pancreatoduodenectomy per primam in order to avoid a misdiagnosis and only 20% of cases are considered candidates for transduodenal ampullectomy or other conservative procedures (18). Therefore, accurate diagnosis is crucial to avoid overtreatment and ensure optimal patient care. However, cases presenting complications such as jaundice, bleeding, pain or suspicion of malignant transformation would rather benefit from surgery with radical intent (4). Meanwhile, uncomplicated cases in which the presence of heterotopic pancreatic tissue at the level of the ampulla of Vater is highly suspected might rather benefit from ampullary resection than from complex surgical procedures such as pancreatoduodenectomy (18,19). One of the first successfully treated cases through a conservative approach was reported in 1983 by Laughlin et al; at that moment the authors reported the case of a patient diagnosed with an aberrant pancreatic islet at the level of the ampulla of Vater which was resected en bloc with the duodenal papilla followed by reimplantation of the common bile duct at the level of the duodenum (20). However, in the

absence of an adequate treatment, heterotopic pancreatic tissue is susceptible to some complications: biliary tract obstruction, bleeding, pancreatitis, pseudocyst formation or even malignization (21,22).

Management of this type of lesion can be debated if the tumor doesn't have clinical impact, but otherwise, complicated cases with jaundice, pain, bleeding and malignancy will have to be operated.

Conclusions

Heterotopic pancreatic islets at the level of the ampulla of Vater represent a rare situation which might predispose to significant challenges in terms of diagnosis and therapeutic strategies. When it comes to the case that we reported, the patient presented the symptoms of a periampullary tumor while the imagistic studies failed to reveal any obvious lesion in order to orientate the diagnostic to an ectopic pancreatic tissue. Therefore, the case was treated as a pancreatic head tumor and was treated accordingly, although the morbidity rate increased significantly. In this respect, we are enabled to conclude that, by raising awareness of this rare entity and its diagnostic challenges, clinicians can be better equipped to identify and manage heterotopic pancreas in the ampulla of Vater or other sites effectively, without increasing the risk of perioperative morbidity.

Author's Contributions

Conceptualization: R.C., B.G.; L.A.; methodology: A.C.; validation: M.H.; M.T.; A.P., G.M.; formal analysis: A.H., A.C.; investigation: C.M., S.P.; C.D.; resources: I.B.; data curation: G.P.G., A.Z.; writing — original draft preparation: R.C.; V.V.; L.P.; writing — review and editing: I.B.; visualization: B.G.; supervision: N.B.; project administration: M.S. All authors have read and agreed to the published version of the manuscript.

Conflicts of Interest

There are no known conflicts of interest in the publication of this article. The manuscript was read and approved by all authors.

Ethical Statement

The study was conducted according to the guidelines of the helsinki declaration.

Informed consent was obtained from the subject involved in the study.

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