Ectopic pancreas located in the major duodenal papilla: case report and review

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The term ectopic pancreas indicates the presence of well-developed and normally organized pancreatic tissue outside the pancreas without anatomic or vascular connections with the pancreas proper. Most occurrences of ectopic pancreas are located in the stomach, duodenum, or jejunum. However ectopic pancreas sometimes can be found in unusual sites such as Meckle diverticulum, gallbladder, umbilicus, mediastinum, and the fallopian tube. Ectopic pancreas rarely produces clinical symptoms but occasionally it can result in symptoms that vary according to its location, size, and the involvement of overlying mucosa. An unusual case of ectopic pancreas in the major duodenal papilla is presented here. The patient presented with vague epigastric pain and bile duct dilatation. Published reports are reviewed with respect to imaging, diagnosis, and management of ectopic pancreas of the papilla.

CASE REPORT

A 59-year-old woman was admitted for the evaluation of intermittent upper abdominal pain and postprandial abdominal fullness of 6 months’ duration. The pain was vague and unrelated to position change. On physical examination there was minimal tenderness in the epigastrium. There was no fever, jaundice, weight loss, or abnormal lymph node enlargement. Chest x-ray and EGD were unremarkable. Abdominal US demonstrated a dilated bile duct (1.3 cm) and dilated intrahepatic ducts. There was no mass identified by US. Laboratory data included a hematocrit of 36.2% (normal 37%-47%) and a white blood cell count of 6800/mm³ (normal 4800-10,800/mm³). Serum amylase was 97 U/L (normal 25-125 U/L), lipase 142 U/L (23-300 U/L), aspartate aminotransferase 25 U/L (10-35 U/L), alanine aminotransferase 38 U/L (5-40 U/L), serum direct bilirubin 0.1 mg/dL (0.1-0.5 mg/dL), serum total...
bilirubin 0.5 mg/dL (0.2-1.0 mg/dL), and alkaline phosphatase 88 U/L (66-220 U/L). Carcinoembryonic antigen was 1.01 ng/mL (normal < 10 ng/mL) and carbohydrate-associated antigen 19-9 (CA 19-9) was 3.84 U/mL (normal < 37 U/mL). ERCP revealed dilation of the bile duct and the main pancreatic duct (Fig. 1). Biopsy specimens were taken from the major duodenal papilla although it looked normal during cannulation (Fig. 2). Histopathologic evaluation disclosed only chronic inflammation. EUS demonstrated a hypoechoic tumor (2.5 cm × 1.3 cm) within the muscularis propria (Fig. 3), and a tentative diagnosis of leiomyoma located in the major duodenal papilla was made. Laparotomy was performed because of the uncertainty as to the nature of the tumor and a concern that it might be malignant. At exploration the pancreas was normal. However the major duodenal papilla had a relatively hard consistency and measured 1.2 cm × 1.2 cm. A local excision of the major duodenal papilla with sphincteroplasty was performed. The postoperative course was uncomplicated. Histologic evaluation revealed an ectopic pancreas with interlacing bundles of smooth muscle in the major duodenal papilla (Fig. 4). The patient remained asymptomatic and US disclosed pneumobilia without bile duct dilation at 6 months after discharge.

DISCUSSION

Most tumors that cause bile duct obstruction are periampullary cancers, including ampullary carcinoma, carcinoma of pancreatic head and distal bile duct cancers. Benign tumors of extrahepatic duct can cause jaundice or abdominal pain, adenomas and papillomas being the most common. Ectopic pancreas located in the major duodenal papilla is unusual with a total of only 8 cases having been reported (Table 1).

The diagnosis of ectopic pancreas is easier to make when it exhibits a characteristic central dimpling. However only the larger size of the major papilla was noted in the reported cases of ectopic pancreas located in the major duodenal papilla and in no case was the diagnosis made before laparotomy. Similarly the surface of the major duodenal papilla was smooth in our patient and the tumor measured 1.2 cm × 1.2 cm in size.

The most common EUS appearance of ectopic pancreas in the GI tract is that of a hypoechoic or of mixed echogenic lesions located in the submucosal layer. EUS features of ectopic pancreas in the major duodenal papilla have not been reported. A hypoechoic tumor in the muscularis propria of the major duodenal papilla was noted in our case. When these features are present on EUS of the major duodenal papilla, ectopic pancreas should be included in the differential diagnosis in addition to leiomyoma.

Although endoscopic sphincterotomy may increase the yield of biopsy in diagnosing tumors involving the major duodenal papilla, it may alter the histologic appearance and induce complications such as pancreatitis, hemorrhage, or perforation. It is not known whether EST carries an increased risk of pancreatitis in patients with ectopic pancreas in the major duodenal papilla or whether it helps in the diagnosis. EUS-
guided fine needle aspiration has been widely and safely used in the diagnosis of gastric submucosal tumors and could be considered in cases of benign-appearing tumors involving the major duodenal papilla.

It is uncertain whether ectopic pancreas caused abdominal discomfort to our patient although she had no jaundice or abdominal complaints after operation. In all 8 reported cases of ectopic pancreas located in the major duodenal papilla the patients presented with abdominal pain and 5 of them had clinical jaundice. Hayes-Jordan et al. pointed out that ectopic pancreas is frequently associated with muscle hypertrophy. However only pancreatic acini and ducts interlacing with smooth muscle bundles were found in our patient and there was no evidence of smooth muscle hypertrophy.

The management of ectopic pancreas remains controversial. DeCastro Barbosa et al. reported that 61% of the 41 cases of ectopic pancreas had symptoms attributable to the ectopic pancreas itself. However, Dolan et al. concluded that the majority of the abdominal discomfort did not arise from ectopic pancreas because the symptom could be controlled as often with medical treatment as with surgical treatment. Surgical excision of the ectopic pancreas was recommended by Jochimsen et al. if symptoms persisted after excluding other coexisting diseases such as peptic ulcer, gastroesophageal reflux, or biliary tract disease. Resection of the pancreatic tissue-bearing area was recommended by Tanaka et al. when ectopic pancreas is encountered coincidentally at operation to avoid the possibility that it might in the future produce symptoms. Asymptomatic patients should be observed if the benign nature of the lesion can be ascertained and excision should be performed if the nature of the lesion is uncertain. In symptomatic patients excision is recommended when other causes can be excluded.

Table 1. Summary of clinical features of 9 patients with ectopic pancreas in the major duodenal papilla

<table>
<thead>
<tr>
<th>Author (reference)</th>
<th>Case No.</th>
<th>Age/Gender</th>
<th>Jaundice</th>
<th>Bile duct dilation</th>
<th>Tumor size</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decastro Barbosa et al. (4)</td>
<td>1</td>
<td>61/F</td>
<td>–</td>
<td>+</td>
<td>2 mm</td>
<td>Excision</td>
</tr>
<tr>
<td>Decastro Barbosa et al. (4)</td>
<td>2</td>
<td>55/M</td>
<td>+</td>
<td>+</td>
<td>12 mm</td>
<td>Autopsy</td>
</tr>
<tr>
<td>Hoelzer (5)</td>
<td>3</td>
<td>54/F</td>
<td>–</td>
<td>+</td>
<td>3 mm</td>
<td>Autopsy</td>
</tr>
<tr>
<td>Varay (6)</td>
<td>4</td>
<td>44/F</td>
<td>–</td>
<td>+</td>
<td>25 mm</td>
<td>Incision</td>
</tr>
<tr>
<td>Pearson (7)</td>
<td>5</td>
<td>43/F</td>
<td>–</td>
<td>+</td>
<td>8 mm</td>
<td>Excision</td>
</tr>
<tr>
<td>Weber et al. (8)</td>
<td>6</td>
<td>46/F</td>
<td>–</td>
<td>+</td>
<td>5 mm</td>
<td>Excision</td>
</tr>
<tr>
<td>Laughlin et al. (9)</td>
<td>7</td>
<td>54/F</td>
<td>–</td>
<td>+</td>
<td>1715-5</td>
<td>Whipple's operation</td>
</tr>
<tr>
<td>Kubota et al. (10)</td>
<td>8</td>
<td>71/M</td>
<td>–</td>
<td>+</td>
<td>12 mm</td>
<td>Surgical ampullectomy</td>
</tr>
<tr>
<td>Chen</td>
<td>9</td>
<td>59/F</td>
<td>–</td>
<td>+</td>
<td></td>
<td></td>
</tr>
</tbody>
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REFERENCES