

Ectopic Pancreas: Usual and Unusual Features

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Abstract. Nine patients with ectopic pancreas in the stomach (8 patients) and duodenum (1 patient) were studied both radiographically and endoscopically. Correct diagnosis was made by radiography in six cases and by endoscopy in seven cases. Masses radiographically larger than 3 cm in diameter were seen in three patients. The incorrect radiographic diagnoses were related to the presence of a large mass in one patient and to the complications of severe bleeding and gastric outlet obstruction in the other two. Endoscopy and radiography are complementary modalities in the diagnosis of ectopic pancreas.

Key words: Ectopic pancreas – Pancreatic rest, symptoms, complications – Radiography – Endoscopy.

Ectopic pancreas, or pancreatic rest, is a relatively rare entity and aberrant pancreatic tissue is usually of no clinical importance. In most patients with ectopic pancreas, endoscopy and radiography lead to the correct diagnosis [1-4]; however, resection is necessary in some cases to exclude neoplasm or to prevent complications related to the pancreatic rest [5, 6]. Recently upper gastrointestinal series were performed in nine patients who, during subsequent endoscopy or surgery, proved to have ectopic pancreas in their stomach or duodenum. Six of these patients were correctly diagnosed from radiographs. In the other three patients with severe upper gastrointestinal symptoms, the radiographic diagnosis was incorrect because of the unusual radiographic appearance of the mass or because of clinical complications.

Materials and Methods

The charts of 70 patients evaluated from 1970 through 1978 with a diagnosis of ectopic pancreas proved by

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autopsy, surgery, or endoscopy were reviewed. Nine of these patients had undergone a UGI series and they form the basis of this report. In each of these nine cases, the symptoms, radiographic findings, and the presence or absence of complications related to the anomaly were reviewed.

Results

The ages of the nine patients ranged from 27 to 72 years (average, 43). There were five men and four women. Only three of the nine patients exhibited marked symptoms: one had severe pain, one hematemesis, and one prolonged nausea and vomiting. Three patients had no UGI symptoms and the ectopic pancreas was detected incidentally (during evaluation for metastatic disease in two patients and diarrhea in the other). The remaining three patients complained of abdominal discomfort. Six patients underwent surgery for removal of the gastric lesion, while one without symptoms and two with abdominal discomfort were either conservatively treated or not treated at all after a small, smooth mass with a central umbilication was endoscopically demonstrated. Of the six surgically treated patients, four had no recurrent symptoms after surgery and the other two had symptoms relieved by antacids.

Table 1 summarizes the radiographic features observed in the nine patients. Six showed a smooth

Table 1. Radiographic presentation

Radiographic features	No. of patients
Size of mass	
small (< 1 cm)	4
medium (1-2.9 cm)	2
large (\geq 3 cm)	3
Umbilication	6
Site of mass	
Antrum of stomach	7
Body of stomach	1
Duodenal bulb	1

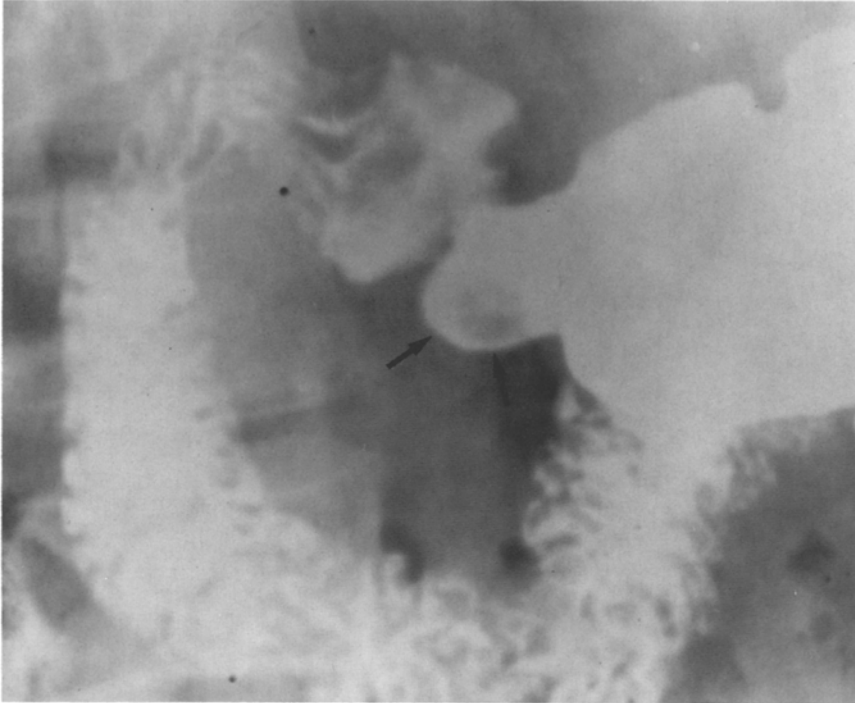


Fig. 1. Smooth round mass (*black arrows*) in antrum close to greater curvature with central collection of contrast material suggesting pancreatic rest

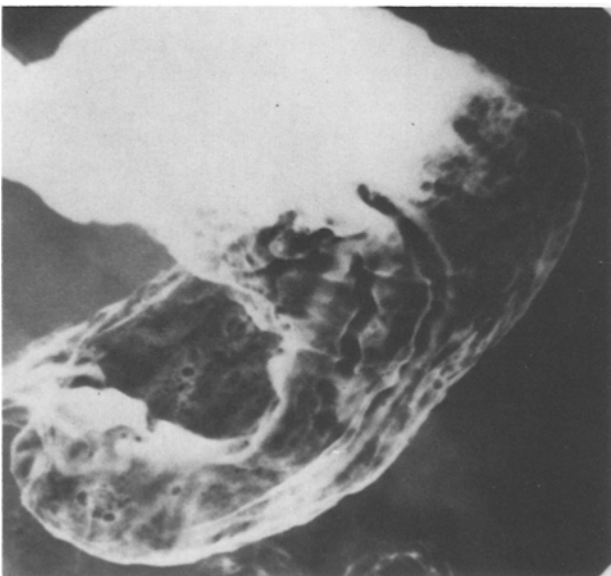


Fig. 2. Large broad-based, lobulated mass in body of stomach with no evidence of central collection of contrast material. Histologic examination showed ectopic pancreas

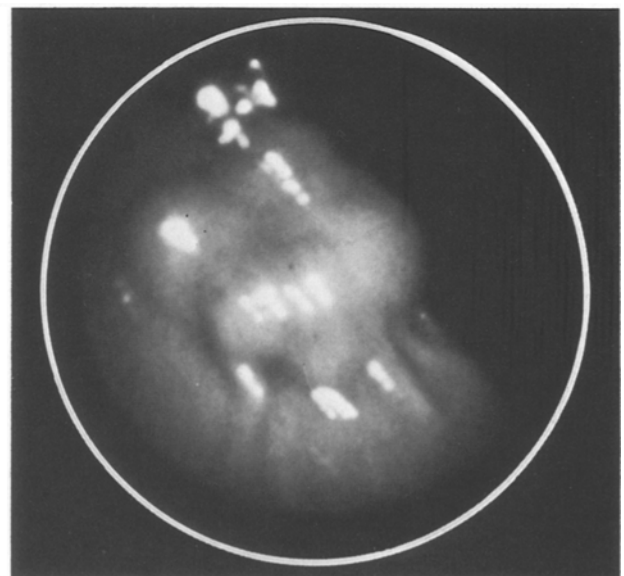


Fig. 3. Endoscopic picture of the large lobulated mass in Fig. 2 with no umbilication

mass with central umbilication (Fig. 1). In all nine patients, none of the superficial biopsies were diagnostically helpful. One small lesion appeared to be pedunculated. In three of the five medium-to-large lesions, umbilication could not be shown. Endoscopy demonstrated the umbilication or duct in the pancreatic rest in seven patients. Four patients had lesions of 1 cm or less in diameter. The largest mass measured

6 cm in diameter and was in the body of the stomach (Fig. 2). Neither radiography nor endoscopy (Fig. 3) demonstrated umbilication in this lesion and it was misinterpreted as a leiomyoma.

Two patients had major complications. One, with a large pancreatic rest in the immediate prepyloric area, had gastric outlet obstruction that radiographically simulated adult hypertrophic pyloric stenosis

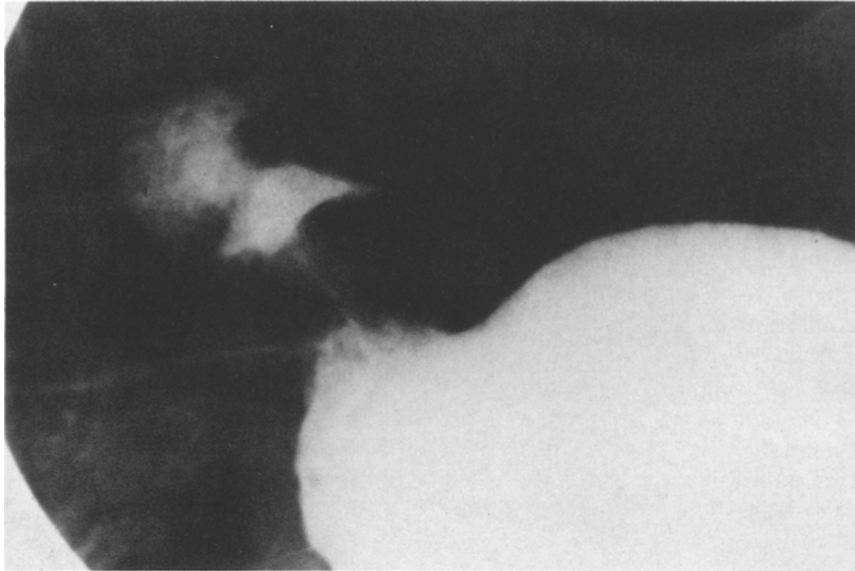


Fig. 4. Radiograph of a 70-year-old woman with severe vomiting shows gastric outlet obstruction with minimal passage of barium contrast material through an elongated pylorus and a "shoulder" deformity of the lesser curve of the antrum mimicking hypertrophic pyloric stenosis. Surgical resection of the distal stomach revealed ectopic pancreas with pancreatitis



Fig. 5. Histologic specimen of the ectopic pancreas seen in Fig. 4 demonstrates dilated ducts (*straight arrows*), acinar tissue (*curved arrows*), and evidence of pancreatitis in the form of interstitial edema with leukocytic infiltrate

(Fig. 4). Endoscopy did not change this diagnosis. This patient was evaluated initially for severe vomiting, and laboratory tests showed an elevated amylase level. The histologic specimens (Fig. 5) of antrum and pylorus showed inflammation, dilated ducts, and cystic areas in nests of pancreatic tissue that were interspersed in the submucosa and muscularis mucosae. The findings were consistent with pancreatitis of the ectopic pancreas with pseudocyst formation.

The other complication was severe upper gastrointestinal bleeding in a hemophiliac. In that case ra-

diographic appearance of a smooth mass with a large central collection of barium was interpreted as a bleeding leiomyoma. Endoscopy showed that the hemorrhage originated from superficial erosions in the mucosa overlying the surgically proved pancreatic rest; a small umbilication could be demonstrated adjacent to the ulcer and the correct diagnosis of pancreatic rest was made.

Histologic examination of specimens from the six patients who underwent resection of the mass revealed that five patients had pancreatic rests with all pan-

creatic cell types present: pancreatic acini, islets of Langerhans, and pancreatic ducts. The sixth patient showed only pancreatic acini and ducts.

Discussion

Ectopic pancreas is usually an incidental finding at surgery or autopsy. The incidence of ectopic pancreas in autopsy series ranges from 1 to 2% [6–8]. However Feldman and Weinberg [9] found an aberrant pancreas in 12.5% of 265 consecutive adults at autopsy. Barbosa et al. [10] found ectopic pancreas in 0.5% of patients undergoing laparotomy. The high incidence in the study of Feldman and Weinberg [9] may be explained by the fact that their investigation was meticulous and detected many lesions smaller than 0.5 cm in diameter. Ectopic pancreatic tissue can be found in patients of any age, but it is most frequently seen in middle-aged patients, as in our series. This is probably because the nodules often are too small to be recognized in younger patients [9]. Ectopic pancreas grows very slowly, and the majority are relatively small even in adults.

Many sites for ectopic pancreas have been reported [5–7, 11–19]. The most common are the stomach (24 to 38%), the duodenum (9 to 36%), and the jejunum (0.5 to 27%) [1,8,9, 20–22]. In the duodenum they occur mainly in the area proximal to the papilla or at the papilla. They are relatively rare in the duodenal bulb (9%). Our series consisted mainly of gastric lesions with only one in the duodenal bulb. The ileum is reported to be involved in 3 to 6% of the patients with pancreatic rests, and Meckel's diverticulum is the site in 2 to 6.5% [1, 8, 10, 20].

Ectopic pancreas generally appears pathologically as a smooth nodule and occasionally as a mass with an irregular surface. It is usually situated in the submucosa and only occasionally expands into the muscularis [6–9]. During endoscopy or by gross inspection of a surgical or autopsy specimen, the mass is broad based, appears to project into the lumen of the stomach or bowel, and is rarely pedunculated. An aberrant pancreas can easily be mistaken for a neoplasm because it appears as an isolated, yellowish mass; however, demonstration of umbilication suggests the proper diagnosis. Often, when the umbilication is not seen, ectopic pancreas can mimic other tumors at endoscopy [6]. In our series, endoscopy showed the umbilication and therefore allowed for a correct diagnosis in seven of the nine patients. Cannulation of a pancreatic nodule during endoscopy has been reported [23]; however, that report is probably anecdotal because most aberrant pancreatic ducts are quite short.

Histologic examination provides the definitive diagnosis. All three types of pancreatic tissue are usually present, but some pancreatic rests demonstrate only one or two types [21, 24]. We found only one patient with tissue limited to pancreatic acini and ducts.

Our cases confirm the radiographic literature, which describes the ectopic pancreas as a small, smooth, round or oval mass with a broad base that, on occasion, appears pedunculated [9, 25]. It most frequently occurs in the antrum within 3–6 cm of the pylorus and is on the greater curvature side of the posterior wall of the stomach. A collection of contrast material in the center of the mass helps to make the diagnosis. This collection represents the barium-filled duct, which may be up to 10 mm long and 5 mm in diameter [26]. This duct or umbilication was seen radiographically in two-thirds of our nine patients – a higher incidence than many reports suggest [22, 25]. That all six of our “classic” uncomplicated cases were diagnosed correctly radiographically and seven cases endoscopically shows how these two modalities play an important role in the presurgical diagnosis of ectopic pancreas. If complications occur or if the ectopic pancreas exhibits unusual features, correct diagnosis may be difficult [9, 27, 28].

Complications can be due to the specific site of the lesion. For example, when the lesion is in the prepyloric area and causes gastric obstruction [29], or when the lesion is in the common bile duct or at the papilla of Vater and obstructs the common bile duct [12, 28, 30]. Complications can also follow pathologic changes within the ectopic pancreas or in the tissue adjacent to or overlying the pancreatic nodule. Changes consisting of pancreatitis [18, 31], pseudocyst formation [32], pancreatic adenoma, and carcinoma of the aberrant pancreas [9, 25, 33, 34] have been reported. Generally, however, complications are rare [9, 35].

One of the two complications in our series occurred in a patient with gastric outlet obstruction. The prepyloric location of the ectopic pancreas led to the obstruction, which finally occurred because pancreatitis and pseudocyst formation developed within the ectopic pancreas. The radiographic, and even endoscopic, findings in such a case often cannot be distinguished from those of hypertrophic pyloric stenosis [29, 36]. We know of no previous description of ectopic pancreas mimicking adult hypertrophic pyloric stenosis in which the ectopic pancreas caused gastric outlet obstruction by means of pancreatitis and pseudocyst formation rather than by presence of the pancreatic rest itself. The second complication was the hemophiliac who had severe bleeding caused by ulceration of the mucosa overlying the ectopic

pancreas. Hemorrhage caused by an ectopic pancreas has occurred [27, 34, 37], but in our patient the bleeding from the anomaly became life-threatening as a result of the associated blood dyscrasia. Both of these cases were misinterpreted radiographically and only histologic examination of the surgical specimen revealed the definite diagnosis. Endoscopic appearance suggested the presence of a pancreatic rest in the hemophiliac.

Both endoscopy and radiography led to misdiagnosis in the patient with the very large mass in the body of the stomach but no evidence of umbilication. This patient demonstrated no complication but had severe pain that may have been caused by the large size of the lesion.

It is inconclusive whether an uncomplicated ectopic pancreas can cause clinical symptoms [8]. A long list can be made of symptoms associated with pancreatic rests. It would include pain, abdominal distress, nausea, and vomiting as the most frequent [1, 9, 38–41]; however, in most cases, ectopic pancreas will not produce these symptoms unless they occupy a particular site or complications arise [25, 38]. Resection of the aberrant pancreas relieved the pain in all four of our patients who had symptoms and underwent surgery. Four, however, is too small a number to make definite conclusions regarding the value of surgery. The remaining two patients in our series, who complained of abdominal discomfort but did not undergo surgery, became asymptomatic after treatment with antacids. Therefore, the symptoms may not necessarily be related to the presence of the pancreatic rest.

If a pancreatic rest is diagnosed radiographically and endoscopically, surgical treatment should be avoided. However, if complications are present, radiography and endoscopy may not lead to a correct diagnosis, and surgery may be necessary.

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