HETEROTOPIC PANCREAS
CAUSING SYMPTOMS

Major KENNETH R. DIRKS
Medical Corps, United States Army

Lieutenant-Colonel T. E., FIELD
M.B.E., M.B., D.T.M. & H., R.A.M.C.
Queen Alexandra Military Hospital, Millbank

Pancreatic tissue outside its usual location and without continuity or vascularization with the pancreas proper may be called heterotopic pancreas. Klob (1859) made the first histological studies of aberrant pancreas, in the stomach in one case and the jejunum in another. Since then nearly 600 cases have been reported at either autopsy or operation. For a long time the condition was viewed as a pathological curiosity. Recently, however, with increasing use of subtotal gastrectomy for peptic ulcer, heterotopic pancreas has been recognized as a cause of clinical symptoms. Within a year we have seen three cases with heterotopic pancreas in the duodenal wall. In two the aberrant tissue was clearly responsible for the patients' complaints, while in the third there was also a healed duodenal ulcer.

A woman of 49 had indigestion for 33 years, flatulence, and intermittent epigastric and right hypochondriac pain. Recently her attacks had increased in severity and frequency. A barium meal radiological examination showed a persistent fleck of barium at the base of the duodenal cap with no tenderness, an inability to distend the cap, and delayed gastric emptying with pyloric stenosis. With a diagnosis of duodenal ulcer she had a subtotal gastrectomy. Although the surgeon was certain that he had resected the offending area of the duodenum, careful examination of the specimen revealed no ulcer, but heterotopic pancreas was present in the duodenal wall. The patient has had no symptoms since her operation.

A man of 40 had intermittent attacks of vomiting and abdominal pain for six years, and three times he had vomited blood. Recently his attacks had become longer and more severe. A barium meal radiological examination showed a deformed duodenal cap with two questionable ulcer craters in the posterior wall and rather rapid gastric emptying. He had subtotal gastrectomy for duodenal ulcer, but the surgeon could not demonstrate one. Careful examination of the resected specimen revealed no ulcer but heterotopic pancreas in the duodenal wall. The patient's only complaint since operation has been occasional anorexia in the early morning.

A woman of 45 had bouts of right subcostal pain for 13 years, recently replaced by attacks of vomiting. A barium meal radiological examination in 1956 had shown "old scars of a duodenal ulcer." Two more recent meals showed a small, tender and slightly deformed duodenal cap. Once there was marked delay in gastric emptying but no ulcer was demonstrated. At operation pyloric stenosis with gross scarring of the first part of the duodenum was found. Heterotopic pancreas was present in the duodenal wall.

Discussion

Anatomy. The aberrant tissue occurs in round, firm, finely-lobulated nodules which may be any shade of yellow from tan to cream. These nodules are 1-6 cm. across and often they present a peculiar and diagnostic alteration of the overlying gastric mucosa consisting of a pseudo-diverticulum (Benner, 1951). About 60 per cent of them are in the submucosa of the affected gut (Hudock et al. 1956). Here the pancreatic acini and ducts tend to form tightly-packed, discrete nodules, which raise the mucosa and project into the lumen. Nodules in the muscularis and subserosa are more diffuse and form ill-defined intramural masses. The histological picture may
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be that of normal pancreas, but often the islets of Langerhans and even the acini are lacking. The most common sites (Allen et al. 1952) are duodenum (32 per cent), stomach (31 per cent), jejunum (22 per cent), and ileum (9 per cent). It has also occurred in the mesentery, omentum, spleen, transverse colon, gall bladder, extrahepatic bile ducts, liver, mediastinum, a Meckel’s diverticulum, and an umbilical fistula. In the duodenum it is usually in a limited area in the second portion, between the major and minor duodenal papillae. Heterotopic pancreas has been found in 0.6 to 13.7 per cent of routine autopsies, depending upon the diligence with which it has been sought, and in roughly one of every 500 operations in the upper abdominal region at the Mayo Clinic (Barbosa et al. 1946). The incidence is highest in the fourth, fifth and sixth decades of life, and is two or three times more frequent in males than in females.

Radiology. Heterotopic pancreas presents radiologically in the duodenum, pylorus, or pre-pyloric region of the stomach as a small nipple-like projection, less than 1.5 cm. in diameter in most cases (Littner & Kirsh 1952). A central dimple or umbilication, representing a mucosal depression at the site of the principal excretory duct of the heterotopic tissue, is unfortunately found in only a few cases, but about 75 per cent show radiological abnormality in the stomach or duodenum, usually a benign polypoid tumour or a filling defect suggesting a peptic ulcer. The latter may be seen even though no ulcer is subsequently found in the excised specimen. When a rounded tumour is observed in the first or second part of the duodenum, it should arouse suspicion of an intramural leiomyoma or a mass of aberrant pancreas. Often, however, the most the radiologist can honestly suggest is a polyp or benign tumour.

A survey of the literature on heterotopic pancreas shows three historical phases (Littner & Kirsh, 1952): first the correct embryology was argued, then the serious pathological alterations in these anomalies were described, and the current phase recognizes the entity as a cause of the “duodenal syndrome” or other gastro-intestinal complaints. It seems that aberrant pancreatic tissue may fall heir to all the diseases of the normal organ. Pancreatitis, hemorrhage, necrosis, cyst formation, benign neoplastic change, malignant degeneration, and hypersecretion of islet cells producing hyperinsulinism, have all been found. Massive gastro-intestinal hemorrhage from heterotopic pancreas (Hudock et al. 1956) has been reported six times. The presence of aberrant pancreatic tissue in the pyloric region and in the duodenum has produced radiological and clinical findings so strikingly similar to those of peptic ulcer as to lead to subtotal gastrectomy in a number of cases. Not only peptic ulcer, but gastritis, cholecystitis, common bile-duct obstruction, and gastric neoplasm may be clinically simulated by aberrant pancreas. The symptoms are probably related to local alteration in gastro-intestinal motility with muscle spasm due to the presence of the heterotopic mass. Inflammation around the heterotopic tissue or prolapse of the mass through the pylorus may both cause pyloric obstruction.

A recent study of 51 cases collected in the Mayo Clinic in 47 years (Martinez et al. 1958) indicates that patients fall into three groups: those in whom the lesion is clinically significant, those in whom it is coincidental with other significant pathology and those in whom it is merely an incidental finding. Twenty-eight (56 per cent) of the Mayo Clinic cases were clinically significant, seven (14 per cent) coincidental, and
16 (32 per cent) incidental. All of the clinically significant patients had abdominal symptoms which were no different from those of the more common types of gastro-duodenal or cholecystic disease. Sixteen of these people had epigastric or sub-costal pain and the remaining twelve complained of epigastric distress. In two of our cases the heterotopia was clinically significant and in the other coincidental with a healed duodenal ulcer and pyloric stenosis.

In the past most cases have been treated by subtotal gastrectomy, usually with a pre-operative diagnosis of peptic ulcer. Such extensive surgery is of course unnecessary, for the uncomplicated condition is cured by local excision. The challenge lies in making the diagnosis by a closely co-ordinated effort of radiologist, surgeon and pathologist. The X-ray picture often suggests a polypoid benign tumour, ideally a nipple-like deformity in the gastric or duodenal wall. In other cases the findings suggest a peptic ulcer, and the observations of the surgeon are of paramount importance. If he finds a yellowish nodular lesion which he suspects of being heterotopic pancreas he should not hesitate to remove it locally and request frozen section by the pathologist. The patient may thus be spared needless surgery.

Summary

Three cases of heterotopic pancreas are described. In two the heterotopia was responsible for gastro-intestinal symptoms simulating peptic ulcer; in the third there were also a healed duodenal ulcer and pyloric stenosis. The macroscopic and radiological appearances are presented with particular reference to operative diagnosis and treatment.

REFERENCES


Up the Navy!

You know the worthy lieutenant in command of a vessel, who had different medicines numbered 1 to 9 for specific diseases of the crew. When he ran out of No. 9 he mixed together number 6 and 3 and administered.

(In a letter from Florence Nightingale dated 1888).
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Kenneth R. Dirks and T. E. Field

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