Royal Australasian College of Surgeons

Duodenal pancreatic heterotopia causing acute pancreatitis with gastric outlet obstruction

Heterotopic pancreas (HP), less accurately known as ectopic, accessory or aberrant pancreas is an isolated, firm, somewhat lobulated, nodule that is anatomically separate from the normal pancreas. It is present in at least 1% of individuals and is most often found in the wall of the stomach, duodenum or a Meckel diverticulum but can be present elsewhere in the small bowel, oesophagus, appendix or other intra-abdominal viscera.[1] In the gastrointestinal tract, HP is most often submucosal rather than subserosal and may extend into the muscularis propria to a variable degree.[2] In children, HP is usually asymptomatic and detected incidentally but may rarely cause intussusception, gastric outlet obstruction or gastrointestinal bleeding.[1] Malignant degeneration in this age group is exceedingly uncommon.[3] We report a boy with heterotopic pancreatitis causing gastric outlet obstruction.

An 8-year old boy presented to hospital with recurrent coffee ground vomiting. As a neonate, he had undergone repair of oesophageal atresia and a tracheo-oesophageal fistula. An upper gastrointestinal endoscopy revealed circumferential erosive oesophagitis in the mid oesophagus and he was treated with oral Omeprazole. Nine days later he returned with upper abdominal pain and vomiting. Laboratory analysis revealed a plasma lipase of 600 U/L and CRP of 60 mg/L. An abdominal CT scan showed focal thickening of the gastric antrum with an adjacent triangular wedge of tissue suggestive of heterotopic pancreas (Fig. 1).

He was initially managed conservatively with intravenous fluids, analgesia and nasogastric drainage. Total parenteral nutrition was started because of persistent large nasogastric losses. A repeat CT scan 1 week later revealed multiple pseudocysts around the porta hepatis and anterior to the liver. The patient had ongoing abdominal pain, persistent large volume gastric losses and continuing raised inflammatory markers and plasma lipase. An MRI scan showed acutely inflamed heterotopic pancreatic tissue anterior to the pancreatic head adjacent to the duodenal bulb. There was an associated circumferential fluid collection around the pylorus with gastric outlet obstruction. The pancreas appeared normal. After 3 weeks of failed conservative management a laparotomy was performed.

Multiple large inflammatory pseudocysts around the liver, pylorus and within the lesser sac were drained. The gallbladder was thick walled and distended. A 3 cm long firm pyramidal wedge of heterotopic pancreas was fused to the serosa of the anterior wall of the first part of the duodenum (Fig. 2). A small pancreatic duct entering the heterotopic pancreas from the duodenum was ligated.

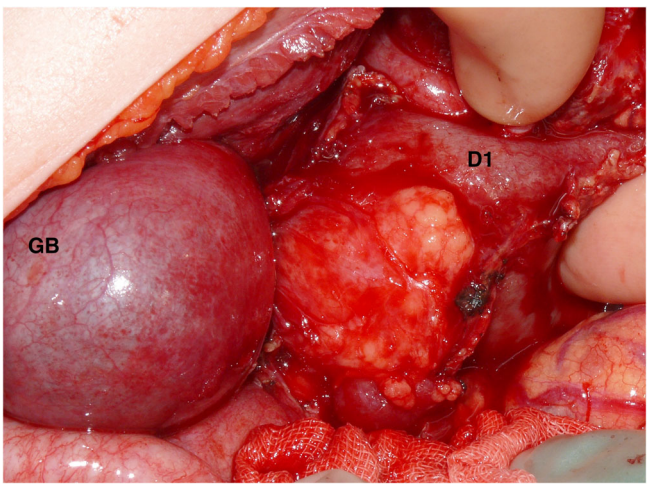

Fig. 2. Operative photograph of the subserosal heterotopic pancreatic mass. D1, first part of duodenum; GB, gallbladder.

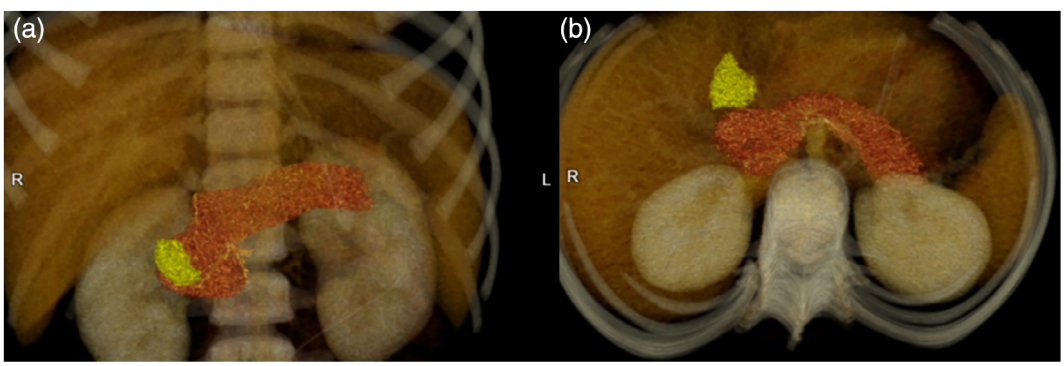

Fig. 1. Axial (a) and coronal (b) CT reconstruction of the heterotopic pancreas (yellow) and the normal native pancreas (red).

The duodenal serosa was repaired and an omental patch applied. He had an uneventful postoperative course and was discharged home 10 days later. Histology revealed pancreatic tissue with normal exocrine and endocrine glands, small ducts and mild fibrosis. The patient remains well 6 months later. His parents kindly gave their permission to report his clinical course.

Pancreatitis within HP tissue in children is very rare. There are only six previous case reports of this complication: two were from HP within an enteric duplication cyst, two were in HP nodules within the jejunal mesentery and one was in the wall of the jejunum.[2,4–6] The sixth report documented heterotopic pancreatitis and gastric outlet obstruction in a neonate who also had infantile hypertrophic pyloric stenosis; the relevance of the HP is not clear as the antropyloric heterotopic tissue was not resected and the patient had no further sequelae during a 2-year follow up.[7]

Our patient also had other unusual features related to the HP. Firstly, the heterotopic pancreatitis caused severe gastric outlet obstruction which resolved after simple resection of the lesion. Secondly, the HP was subserosal rather than submucosal. Thirdly, he had a history of repaired oesophageal atresia. However, it is worth emphasizing that up to 40% of children with a history of oesophageal atresia have evidence of asymptomatic gastric HP on endoscopy.[1]

The pathogenesis of HP is uncertain but the most popular theory is that pancreatic tissue from the lateral buds of the developing dorsal or ventral pancreas becomes attached to an adjacent organ and then detaches from the pancreas. Heterotopic pancreas may contain any of the components of normal pancreatic tissue, namely acini, ducts and islets of Langerhans; all elements were present in the resected specimen in our patient. What triggered the onset of pancreatitis in our patient with HP who had no previous symptoms to suggest similar episodes is unknown.

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Author contributions

William Fleischl: Data curation; writing – original draft. **Ray Li:** Data curation. **Gianluca Valsenti:** Data curation; investigation. **Tim Foster:** Investigation. **Mark D. Stringer:** Conceptualization; project administration; supervision; writing – review and editing.

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