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CASE REPORT

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Combined ileal heterotopic pancreatic and gastric tissues causing ileocolic intussusception in an infant

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Abstract Pathological intussusception is rare in infancy with Meckel's diverticulum being the most common lesion. Isolated heterotopic pancreatic tissue in the serosa of the ileum causing intussusception is extremely rare. We report a case of ileal heterotopic pancreatic tissue associated with ectopic gastric mucosa causing ileocolic intussusception in an infant, with review of the literature.

Keywords Heterotopic pancreas · Ectopic gastric mucosa · Intussusception · Ileal · Pediatric

Introduction

Most intussusceptions that occur in infancy are idiopathic in origin [1]. In a small proportion of cases a pathological lead point is identified as the cause of an intussusception, with Meckel's diverticulum being the most common lesion [2–4]. The incidence of intestinal ectopia in a Meckel's diverticulum is approximately 30–50% [2]. Isolated heterotopic pancreatic tissue in the ileum causing intussusception is extremely rare [2]. Combination of ileal serosal heterotopic pancreatic tissue and an ectopic gastric mucosa as a cause of intussusception is even more of a rarity [1]. We report a case of combined ileal serosal heterotopic pancreatic and mucosal gastric tissues causing an ileocolic intussusception in an infant.

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Case report

A 6-month-old, twin boy, presented with a 1-day history of crying, irritability, pallor, and drawing up his legs. He was born at 35 weeks gestation with a birth weight of 2.5 kg and had been previously well. He had a loose bowel motion (no blood) 12 h prior to presentation and no vomiting.

Physical examination revealed a pale and tachycardic child who looked unwell. The abdomen was soft and non-distended with a palpable mass in the left upper quadrant. There were no peritoneal signs, bowel sounds were present, and rectal examination was unremarkable.

An ultrasound scan was carried out, prior to surgical consultation, which confirmed the diagnosis of intussusception (Fig. 1). Attempted air enema reduction was unsuccessful.

At laparotomy an ileocolic intussusception, with extensive necrotic bowel, and reactive mesenteric lymphadenopathy was found. A limited right hemicolectomy was performed.

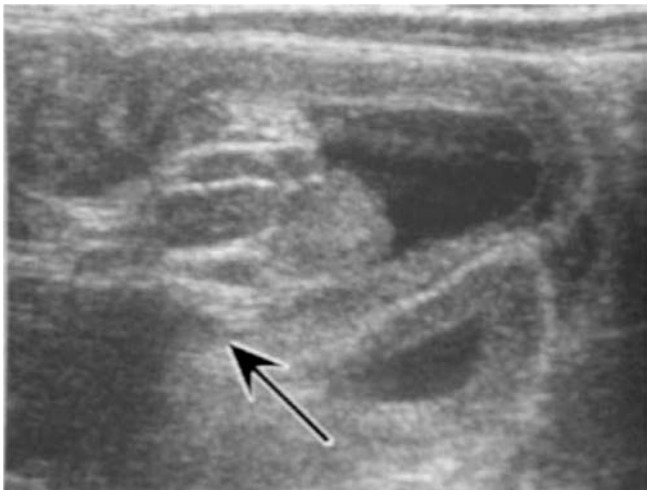

Fig. 1 Ultrasound scan showing the intussusception with possible pathological lead point (*arrow*)

The patient's post-operative recovery was uneventful and he was discharged on the fifth post-operative day.

The specimen consisted of ascending colon and ileum, with 120 mm of ileum intussuscepted through the ileo-caecal valve into the cecum. The distal 100 mm of the intussuscepted ileum was dark, haemorrhagic, and appeared non-viable close to the tip of the intussusception. The lead point of the intussusception was a firm pale nodule (6×5×5 mm^3), which appeared to be within the wall.

On histology the nodule was heterotopic exocrine and endocrine pancreas with ducts present. It was within the serosa and the outer coat of the muscularis propria. No duct was seen connecting with the mucosa. The mucosa in this area though necrotic and haemorrhagic was thick with absent villous outline compared with adjacent mucosa. The viable glandular epithelium in this area was also different and consisted of mucous cells and rounded cells with pink cytoplasm suggestive of parietal cells. The overall appearance was consistent with heterotopic gastric mucosa (Fig. 2a, b).

Discussion

Heterotopic pancreas can be defined as pancreatic tissue lacking anatomic and vascular continuity with the main body of the pancreas [2, 5]. Heterotopic pancreas has an estimated occurrence of 1 in 500 upper abdominal operations and has been reported to be present in 0.6–5.6% of autopsy cases [6]. It is rarely detected in children [2].

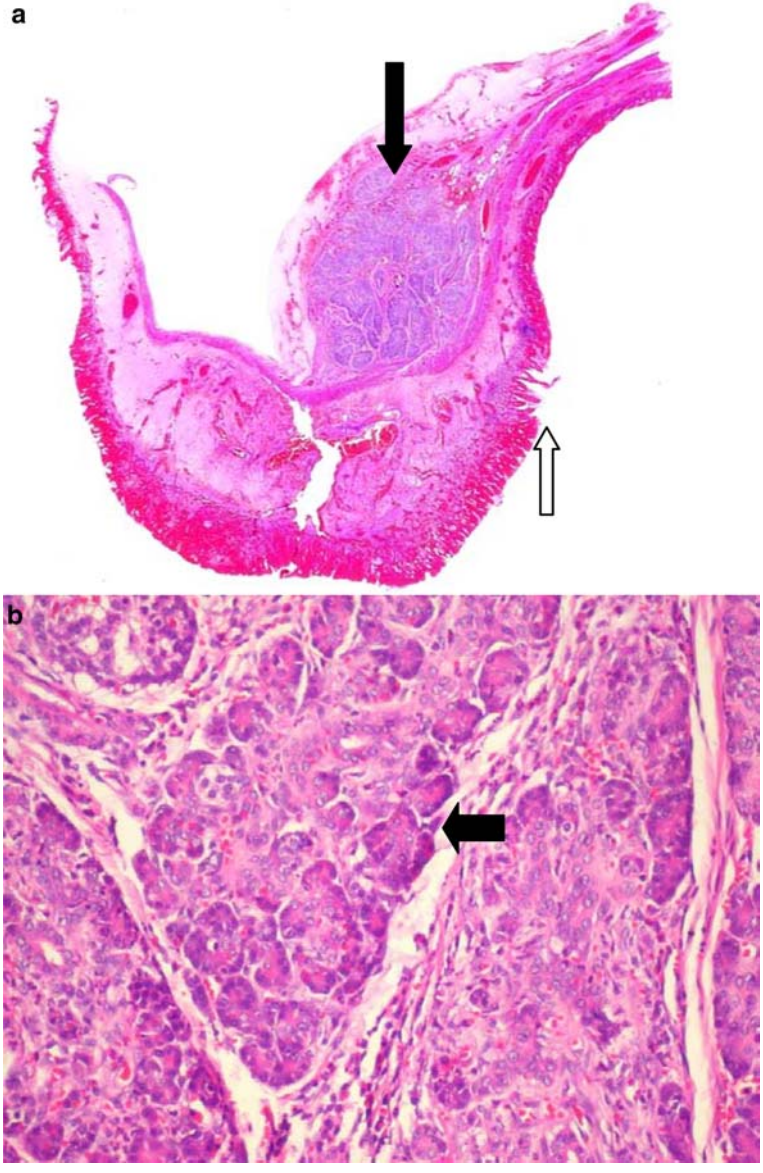

Fig. 2 **a** Histopathology showing firm pale nodule (6×5×5 mm^3) within the serosa and the outer coat of muscularis propria at the tip of intussusception, the lead point is heterotopic pancreas in the serosa (*black arrow*) associated with ectopic gastric mucosa (*white arrow*). **b** Slide showing pancreatic tissue (*arrow*) within the ileal serosa

The most common locations of heterotopic pancreas are the stomach, duodenum, and jejunum [6, 7]. Isolated pancreatic tissue in the ileum is rare, mainly asymptomatic and usually discovered incidentally [8]. Multiple lesions are exceptional [2].

Studies have shown that heterotopic pancreas can produce clinical symptoms. Symptoms include epigastric pain, haemorrhage, weight loss, nausea, and vomiting. Reported complications include malignant change, cyst formation, pancreatitis, insulinoma, and intussusception [5].

It has been reported that two-thirds of isolated ileal heterotopic pancreas cases are found incidentally at the time of surgery [9]. Correct pre-operative diagnosis of heterotopic pancreas is not made, even in symptomatic patients, with the use of barium enema, ultrasound scan, computer tomography, or laparoscopy [9]. It should be suspected with intussusception in extremes of age, 3 months or less or greater than 2 years, and in cases of chronic intermittent, early recurrent, or double intussusception [2, 10, 11].

Symptomatic isolated ileal heterotopic pancreas should be managed by segmental resection, open or laparoscopic techniques. Incidental findings should be biopsied and sent for frozen section and resection or excision carried out [2].

There are a number of theories of origin with respect to heterotopic tissues. Skandelakis et al. suggested that metaplasia of pluripotential endodermal cells of the embryonic foregut as an origin for heterotopic tissues. This may provide an explanation for the occasional report of an unusual site such as the fallopian tube [12, 13]. Others have suggested that separation of pancreatic tissue occurs during embryonic rotation and fusion of the ventral and dorsal pancreatic buds [5]. Abel et al. suggested heterotopic pancreas, especially when associated with gastric mucosa, represents a vitellointestinal tract remnant, similar to a Meckel's diverticulum without the diverticulum [1]. Nonetheless, all above hypotheses have not been tested yet.

On review of the English language literature the first reported case of isolated ileal heterotopic pancreas causing intussusception was by Barbosa et al. in 1946. This was followed by few case reports in paediatric and adult literatures [2, 7–9, 11, 14–19].

There have only been three cases reported in the English literature to date, of intussusception caused by combined ileal heterotopic pancreatic and gastric tissue. Iucthman et al. reported the first case in a 15-year-old boy with intermittent ileocolic intussusception [10]. Abel et al. reported the others in two 16-month-old boys with ileoileal and ileocolic intussusceptions, respectively, where the heterotopic pancreas was located in the ileal serosal tissue in both cases.

Our case is similar to Abel et al. in regard to the location of heterotopic pancreatic tissue within the serosa associated with ectopic gastric mucosa. However, the age of presentation was younger.

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