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Double ileoileal intussusception caused by a giant polypoid mass of heterotopic pancreas in a child

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Abstract Heterotopic pancreatic tissue has been found in several abdominal and intrathoracic locations. In the ileum, it is a rare, usually asymptomatic, incidental finding.

Conclusion A unique case of a recurrent ileoileal intussusception in an 11-year-old girl is presented caused by a giant polypoid mass composed of ectopic pancreatic tissue that remained undetected during several diagnostic tests during two previous admissions and laparoscopic abdominal exploration.

Key words Heterotopic pancreas · Intussusception · Ileal polyp · Paediatric

Introduction

Normally, heterotopic pancreatic tissue remains asymptomatic. However, depending on location, size, physiologic activity and inflammatory or neoplastic complications clinicians may be confronted with diversified clinical manifestations.

Case report

An 11-year-old girl was referred to our clinic with a 1-month history of recurrent 'crampy' abdominal pain. On admission, she presented with a tender mass at McBurney's point which was sonographically suspicious for an abscess. However, all laboratory studies were normal and the mass disappeared after an enema. The moderate abdominal pain did not improve with conservative measures. Exploratory laparoscopy revealed caecal adhesions to the lateral abdominal wall and a suspected inflamed appendix which was removed, otherwise no specific findings. Histology showed a scarred and obliterated appendix. Two weeks later she was admitted again with abdominal pain. At examination, her abdomen was soft with, again, a slightly tender mass in her lower abdomen, which was interpreted as hard stool. Her presumable faecal impaction resolved with food rest and enema treatments.

Four months later she was referred by her gynaecologist with another mild episode of 'crampy' abdominal pain. On questioning, she admitted that her abdominal pain had recurred regularly after her previous hospital stay. Physical examination revealed a palpable tender mass in the right lower quadrant of the abdomen and ultrasound demonstrated a characteristic 'target sign' suggesting intussusception. All laboratory investigations were normal. At laparotomy, the suspected intussusception was located and delivered through a midline incision. A double ileoileal intussusception was discovered about 40 cm before the ileocaecal valve and easily reduced. As the intussusceptum, a walnut-sized thickening at the top of a 6 cm long stalked polyp, thick as an index finger, could be palpated through the ileal wall (Fig. 1). The mesenterium in that region was considerably indurated. Consecutively, the small segment of the ileum containing the polyp was resected and an end-to-end anastomosis was performed. The girl remains asymptomatic 1 year later.

Microscopic examination showed an approximately 2.5 cm nodule of heterotopic pancreatic tissue located within the splintered muscular layer of the ileal wall at the base and inside the pedunculated polypoid mass. The nodule was composed of regular pancreatic tissue including islets of Langerhans. The prominent ileal mucosa showed extensive ulceration with bleeding and rough fibrosis of the ectopic pancreatic tissue, presumably as a consequence of the chronic mechanical alteration caused by peristaltic waves and recurrent intussusception.

Discussion

In a comprehensive review of 471 cases, the most frequent sites of pancreatic heterotopia were the stomach (25.5%), the duodenum (22.7%), the jejunum (15%) and Meckel diverticulum (5.3%) [1]. To explain the origin of

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Fig. 1 Intra-operative findings after reduction of the double ileoileal intussusception. The paediatric surgeon's hands demonstrate the contour of the polyp through the ileal wall. Note the oedematous, previously invaginated, ileum

this developmental anomaly, Skandalakis et al. [10] encouraged the hypothesis of metaplasia of pluripotential endodermal cells of the embryonic foregut which might explain occasional reports on unusual sites such as the fallopian tube [7]. However, no universally acceptable theory has yet been found. The histological appearance of ectopic pancreatic tissue is usually similar to that of normally situated pancreas and it might be affected by the same pathological conditions. Its clinical significance depends on its location, size, physiological activity and complications. Patients with presumed pancreatic heterotopia may present with various clinical features as abdominal pain, anaemia, melaena, weight loss, haematemesis or gastric outlet syndrome [3]. Predominantly, the ectopic tissue becomes clinically symptomatic and radiologically detectable when it is located in the stomach or duodenum and its size exceeds 1 cm [4, 6]. When becoming symptomatic in the small intestine, pancreatic tissue frequently causes intussusception with consecutive bowel obstruction [2, 5].

In the presented case, the ileal intussusception was intermittent and recurrent which was supported by the histology and the periodic target sign on ultrasound. The initial sonographic and physical findings (tender mass) resolved after an enema which presumably reduced the intussusception, mimicking constipation as the cause of abdominal pain. Macroscopically, the stool was never suspicious for blood, however, testing for occult blood was not done. The heterotopic pancreatic polyp functioning as a leading point for the intussusception was not discovered at exploratory laparoscopy although the pathology was within the ileal part which should be explored for Meckel diverticulum (up to 2 ft from the ileocaecal valve). The reliance in evaluating the small bowel by laparoscopic means may be generally overrated as this procedure is highly dependent on the experience of the paediatric surgeon. Here, the clinical symptoms primarily imitated more regular causes of moderate recurrent abdominal pain such as constipation and chronic appendicitis leading to the delayed correct diagnosis and treatment. Moen and Mack [8] even needed two laparotomies to discover the cause of an intussusception in an adult.

Depending on its location, local excision (e.g. in the stomach) or segmental resection with restoration of the continuity by an anastomosis (e.g. in the alimentary tract) has been shown to be a safe and adequate treatment [6, 9].

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