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Intussusception Secondary to Isolated Heterotopic Pancreas of Meckel's Diverticulum

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Abstract

Intussusception in the pediatric age group is usually idiopathic in origin, and in a small number of patients ranging from 2% to 12%, a pathological lesion as a leading point is identifiable. Of the variety of pathologcial lesions identified as leading points for intussusception, Meckel's diverticulum is the most common but very rarely, isolated heterotopic pancreas is found within it.This is a case report of intussusception in an infant caused by an isolated heterotopic pancreas of the Meckel's diverticulum. The literature on the subject is also reviewed.

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Introduction

Meckel described the congenital diverticulum that bears his name in 1809, and it has remained something of a surgical curiosity ever since. It is present in approximately 2% of the population, but only some 4% of these became symptomatic[1]. When symptoms do arise, they are nearly always the result of inflammation, haemorrhage or intestinal obstruction. Obstruction is usually due to the congenital mesodiverticular band but occasionally results from intussusception. Between 4 and 14% of the complications of Meckel's diverticulum can be attributed to intussusception, which generally produces small bowel obstruction in infancy[2-5].

Heterotopic pancreatic tissue consists of normally differentiated pancreatic tissue without a real anatomic and vascular connection to the pancreas[1,2]. It is a developmental abnormality occurring when primitive cells within the wall of the gastrointestinal (GI) tract differentiate into pancreatic tissue.[1] Ectopic tissue is usually discovered in the submucosa of the upper gastrointestinal (GI) tract, including the stomach, duodenum and jejunum, or within a Meckel's diverticulum. It may contain ectopic gastric, intestinal or pancreatic tissues. Very few ectopic pancreatic tissues produce symptoms, with most cases typically found incidentally at surgery or on autopsy. So,Multiple heterotopic pancreatic tissues and Meckel's diverticulum have rarely been reported[2,3]. But they can be a source of significant morbidity[4] and more frequently cause diagnostic confusion.[2]They are discovered at all ages and there is no sex or race predilection.[1]

Ectopic pancreas is mostly distributed in the upper GI tract with 70%–90% of these lesions occurring in the stomach, duodenum and jejunum.[3,5] The most common congenital anomaly in the gastric antrum, it may also be found in a Meckel's diverticulum, ileum, liver, biliary tract, spleen, omentum or mesentery.[3]

A small bowel follow-through, esophagogastroduodenoscopy (EGD), endoscopic

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ultrasonography (EUS), or computed tomography (CT) scan of the abdomen using contrast have been used with varying degrees of success in diagnosing ectopic pancreas.[3,5,6] Surgical resection is necessary in some cases to exclude neoplasm or to prevent complications.[4]

Here we outline a case of intussuscepting diverticula presenting with overt small bowel obstruction in a 10-month-old male infant with heterotopic pancreatic tissue.

Case Report

A 10-month-old male infant was admitted to Rajshahi Medical College hospital because of abdominal pain, fever and vomiting oftwo days duration. The infant had diarrhea the day prior to admission, and while in hospital, he had passed one bloody stool. Clinically, he was mildly dehydrated, and no masses could be felt on abdominal examination. The possibility of intussuscpetion was raised, and this was confirmed by an abdominal ultrasound. A contrast enema revealed intussusception as far as the splenic flexure.

The patient had a laparotomy through a right supra umbilical transverse incision. The intussusception was reduced manually, and while examining the small intestine, a small diverticulum was found in the distal ileum. This was excised via wedge incision and closed transversely. Postoperatively, the patient did well and was discharged home on the fifth postoperative day.

Figure 1: Meckel**'**s diverticulum causing intussusception

Histology of the resected diverticulum revealed a 1 cm polypoid tissue covered with a flat, severely inflamed and ulcerated intestinal mucosa. In the underlying stroma were abundant glandular ductal structures resembling those in the pancreas and surrounded by slightly irregular thin-walled blood vessels. Among the glandular structures were streaks of smooth muscle tissue as well as lymphatic hyperplasia at the margins.

Discussion

In 1946, Barbosa defined pancreatic heterotopia as the presence of pancreatic tissue outside its usual location and without anatomic relation to the pancreas proper, either by physical continuity or shared vascularization.[7] Thirty years later, Ravitch thought that nonulcerated nodules of ectopic pancreatic tissue located in the stomach or the duodenum may frequently be the basis of ulcer-like symptoms relieved by surgical removal.[4,8]

Theories of pathogenesis include a transplantation or misplacement of embryonic tissue, which develops into mature elements, and pancreatic metaplasia of endodermal tissues during embryonic life.

When pancreatic tissue (including acini, ducts and islets) predominate, the masses are called heterotopic or ectopic pancreas.[1,4] Less frequently, the ductal type of tissue (consisting of ducts of the pancreaticobiliary type surrounded by hypertrophic smooth-muscle bundles) predominates.[1]

The distribution of ectopic pancreas has an embryologic basis.[9] The pancreas arises from dorsal and ventral pancreatic buds. Initially, the dorsal primordium arises from the dorsal aspect of the duodenum; the ventral bud arises slightly later. The dorsal bud is larger and grows faster than the ventral bud. The dorsal pancreatic bud gives rise to the body and tail of the pancreas, whereas the ventral bud gives rise to the uncinate process and part of the head of pancreas. The ventral pancreas is carried away from duodenum by elongation of the hepatic primordium. The ectopic rests drop off from the dorsal primordium and develop in the distal part of the stomach and proximal part of the duodenum.[4,7]

Meckel's diverticulum was first described by Fabricus Hildanus in 1598 [18], and was later named

after the German anatomist, Johann Friedrich Meckel, who described its embryological origin in 1809 [19] It is the vestigial remnant of the vitello-intestinal duct, which acts as a communicating tract between the embryonic yolk sac and its primitive mid-gut in the first few weeks of development. Failure of complete obliteration of the vitello-intestinal tract results in a variety of congenital defects, of which Meckel's diverticulum is the commonest anomaly[18].

Patients with Meckel's are usually asymptomatic, and the diverticulae are invariably discovered incidentally at autopsy, laparotomy or laparoscopy. These patients have a 2–4% lifetime risk of developing complications from it [18,19,20]. Complications from Meckel's usually arise from its underlying mucosa, 50% of which are ectopic mucosae such as gastric mucosa (17.9% – 40%), pancreatic tissue (5–16%), and less commonly, duodenal, colonic and biliary tissue [18,19]. The complications are commonly intestinal obstruction, intussusception, inflammation, haemorrhage and less commonly, perforation, herniation and neoplasm[18,19].

Heterotopic pancreatic tissue itself is uncommon, with reported frequency between 0.55 to 13.7% [22]. It is the presence of pancreatic tissue which lacks anatomical and vascular continuity with the pancreas. It is usually found in the stomach, duodenum and upper part of jejunum, less commonly in the Meckel's, ileum, biliary system, and even spleen. Similar to Meckel's diverticulum, ectopic pancreatic tissues are usually asymptomatic and are found incidentally; they too can occasionally cause symptoms such as bleeding, inflammation, abdominal pain and rarely malignant changes.

The incidence of a localized pathological leading point for intussusception varies from 2%-12% in large series. Ong and Beasely in a review of 630 episodes of intussusception found a pathological leading point in 60 episodes (9.5%)[4]. Issa et al. found a leading point in 8.2% of 233 patients with 244 intussusceptions[6], while Ein found 31 patients (5.5.%) with a leading point among 569 children with intussusception[2]. The variety of pathological lesions identified as leading points in childhood intussusception include Meckel's diverticulum, which is the most common, intestinal polyps, duplication cysts, lymphoma and lymphosarcoma[1-3,4,7]. In a large series of 60 patients with pathological leading points, Meckel's diverticulum was the cause in 27 (45%) of them[4]. Puri and Guiney reviewed 292 consecutive children with intussusception, and 27 of them had identifiable intestinal lesions as a leading point. Among these, Meckel's diverticulum was the cause in 13 (48%) of them, while in 10 cases, intussusception was caused by a tumor of the small intestine[7]. In rare isolated instances, heterotopic intussusception.[5,7-13]

Histologically heterotopic pancreas is classified into 3 types: type 1 (all elements of the normal pancreatic tissue are present); type II (pancreatic tissue without islet cells); and type III (pancreatic ducts only) [9-15]. Our patient had type II heterotopic pancreas. Although, most patients with heterotopic pancreas are asymptomatic, various changes including pancreatitis, pseudocyst formation, inflammation, abscess formation, pancreatic adenoma and carcinoma have been reported in heterotopic pancreas[14-17]. Heterotopic pancreas in the ileum is rare, and when seen, it is usually associated with Meckel's diverticulum.

Isolated heterotopic pancreas of the ileum on the other hand is very rare, usually asymptomatic and discovered incidentally during surgery for other conditions, and very rarely as a leading point for intussusception[5,8,10-12]. In the series of 292 consecutive children with intussusception by Puri and Guiney, none of the 27 patients identified with leading points had isolated heterotopic pancreas[7]. In the review of 630 episodes of intussusception by Ong and Beasley, none of the 60 patients with pathological leading had isolated heterotopic pancreas[4]. In a series of 134 children with intussusception, Hamada et al. found (5.9%)[8] with intestinal lesions as leading points with isolated heterotopic pancreas of the ileum[5]. Pang et al. reviewed 32 histologically proven cases of heterotopic pancreas, of which 14 (44%) were

symptomatic and 18 (56%) were asymptomatic or incidental. Six of the 14 symptomatic cases were in the ileum, and 4 caused intussusception, 2 in Meckel's diverticulum, 1 in a duplication cyst, and 1 as an isolated heterotopic pancreas of the ileum[18]. In an extensive review of the literature, we found only nine cases of isolated heterotopic pancreas of the ileum as a leading point for intussusception in children[5,8,10-12,18]. Their ages ranged from 2 months to 11 years, and in all except two, the intussusception was ileo-ileal, two of which were recurrent intussusception. Whereas idiopathic intussusception is usually seen in infants between 3 and 12 months of age, intussusceptions secondary to a pathological lesion on the other hand have a different age distribution.

The relative frequency of a pathological lesion causing intussusception increases with age, but the first year remains the period with the highest frequency[2,4]. Six of the nine children with isolated heterotopic pancreas of the ileum causing intussusception were less than two years of age . Although there are a few instances in which an intussusception secondary to a leading point has been successfully reduced, most intussusceptions due to a pathological lead point are irreducible by barium enema[2]. With recent advances in ultrasonography, it is now possible to diagnose ileo-ileal intussusceptions pre-operatively, and to avoid delays, let patients undergo open reduction. It is also important to recognize heterotopic pancreas within the wall of the ileum after manual reduction of the intussusception by palpating the bowel. This is necessary for those patients with submucosal location of the heterotopic pancreas where the tumor may not be obvious from the outside, like it was in our patient. Simple excision is the treatment of choice. This will avoid recurrence of the intussusception as well as the risk of its subsequent clinical sequale.

The suspicion of a Meckel's is however often not thought of at the initial stage of patient management, and the diagnosis of Meckel's is quite challenging and it is not infrequently overlooked on radiological imaging unless one is actively looking for it.

Conclusion

Although Meckel's and its complications are not common, the possibility of a Meckel's diverticulum and its potential complications should be considered when faced with a common gastrointestinal complaint and negative initial investigations. When found incidentally at laparotomy or laparoscopy, it should be carefully examined for any gross macroscopic abnormality and resection should be considered, especially in young male patients who are more likely to develop complications from it or patients who might have had previous symptoms attributable to it.

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