Double intussusception with a carcinoid tumor as the lead point in a Meckel's diverticulum

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The authors report the rare occurrence of an intussusception of a Meckel's diverticulum, which was also intussuscepted and found to contain a carcinoid tumor. The tumor may have been the lead point of the intussusception of the diverticulum into itself, and the intussuscepted Meckel's diverticulum may have functioned as the lead point in the intussusception of the ileum. The authors discuss this case in the context of a literature review.

• In 1598, Fabricius Hildanus described a small diverticulum. The anatomical and embryological basis for this congenital gastrointestinal anomaly, Meckel's diverticulum, was elucidated by Johann Friedrich Meckel in 1809.[1,2] Intussusception caused by a Meckel's diverticulum was not reported until nearly a century later, in 1898.[3] The understanding of this condition continued to grow in the early 1900s, with the discovery of ectopic gastric mucosa by Salzer and associated ulceration of the ileum by Deetz.[4-6] In addition, Gramen described a clinical picture of Meckel's diverticulum similar to appendicitis.[7]

Oberndorfer described the first Meckel's carcinoid tumor.[8] Carcinoid tumors in Meckel's diverticula were eventually found to originate from the Kulchitsky cells in Lieberkühn's crypts.[9] Since that discovery in the early 1900s, more than 100 cases of a carcinoid tumor in a Meckel's diverticulum have been reported in the literature. We provide a case report of a 14-year-old boy who had abdominal pain, which was diagnosed as perforated appendicitis. On laparotomy, however, he was found to have an intussusception of a Meckel's diverticulum. The Meckel's diverticulum was itself intussuscepted and contained both ectopic gastric mucosa and a carcinoid tumor.

The tumor may have been the lead point of the intussusception of the diverticulum.

Case report

A 14-year-old boy had a 9-day history of intermittent diffuse abdominal pain. The pain was nonradiating and described as burning. It became increasingly severe and was accompanied by nausea and eight episodes of vomiting on the day he went to the hospital. The patient had not had a bowel movement in 2 days. He did not have diarrhea, fever, rectal bleeding, or viral symptoms. He also had not recently used antibiotics or traveled.

Approximately 1 year earlier, the patient had been treated for several episodes of abdominal pain by a pediatric gastroenterologist. A diagnosis of irritable bowel syndrome was made and he was treated with dicyclomine. He stated, however, that the current pain was constant, unlike that experienced in the past, which usually resolved within a few hours. His surgical history was significant only for repair of a hydrocele at 1 year of age.

The patient was mildly hypertensive and had a temperature of 36.7°C. The physical examination showed a soft, nondistended abdomen, with hypoactive bowel sounds. On palpation, he was tender in all quadrants, but more so in the right lower quad-

rant. There was guarding but no rebound tenderness, masses, or hernias. No masses were found on rectal examination, and results of guaiac testing of the stool were negative. The findings from the rest of the physical examination were normal. The white blood cell count was 14.1 g/dL with a left shift, and the hematocrit was 49%. The alkaline phosphatase level was 330 U/L. All other blood chemistry results were normal.

Acute appendicitis was suspected. On computed tomography (CT) scanning, the oral contrast agent did not reach the cecum, thus the appendix could not be visualized; however, there appeared to be a multiloculated abscess in the pelvis (Figure 1). It was thought that this abscess most likely resulted from an appendiceal rupture.

During an emergency exploratory laparotomy, the abdomen was free of purulence. Exploration of the pelvis showed an edematous, dilated loop of intussuscepted bowel. The intussusception showed signs of full-thickness necrosis and was irreducible. A Meckel's diverticulum, also intussuscepted, was adjacent to the small bowel intussusception (Figure 2). A 58-cm section of small bowel was excised, and the viable ends were anastomosed. The appendix appeared normal and was also excised.

The pathology examination showed an intussusception of the small bowel segment with mucosal necrosis and transmural hemorrhage. The Meckel's diverticulum measured 4.5 x 3 x 3 cm, the end of which was also intussuscepted and contained several focal areas of gastric mucosa. A 2.5-mm carcinoid tumor was present in the tip of the Meckel's diverticulum. Histologic analysis, including neuroendocrine staining, confirmed the diagnosis of a carcinoid tumor (Figure 3). The patient recovered without incident and was discharged to his home on postoperative day 2.

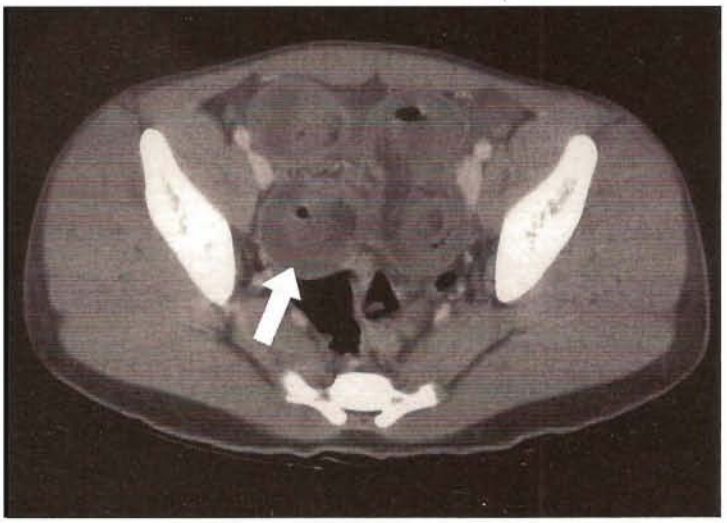

Figure 1—CT scan of the pelvis showing the characteristic target lesion of the intussusception (arrow), which was originally thought to be a multiloculated abscess.

Discussion

Meckel's diverticulum is the most common congenital anomaly of the gastrointestinal tract, with an incidence of about 2% in the general population.[10] It results from an incomplete obliteration of the vitelline (omphalomesenteric) duct, which accounts for 90% of omphalomesenteric duct abnormalities. The omphalomesenteric duct provides the embryo with nutrients from the yolk sac. Normally, during week 8 of gestation, the duct is obliterated and nutrients are supplied to the fetus via the placenta. When this obliteration does not occur, a true diverticulum containing all layers of the gastrointestinal wall results. It is generally found within 2 feet of the ileocecal valve.[3]

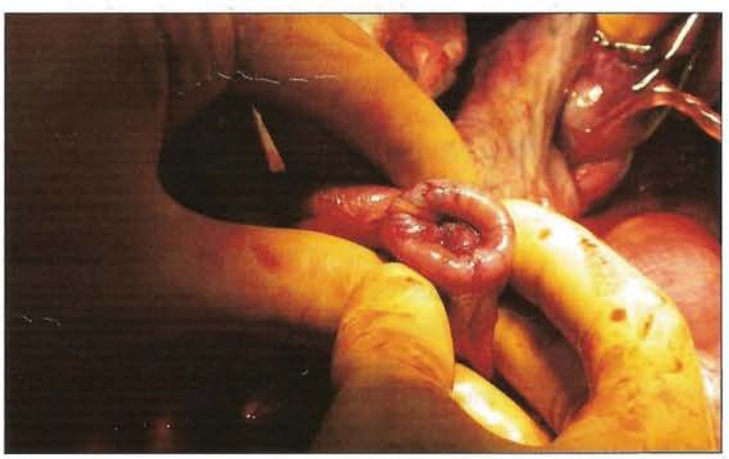

Figure 2—Intraoperative photograph showing the intussuscepted Meckel's diverticulum, which was adjacent to the intussuscepted portion of the small bowel.

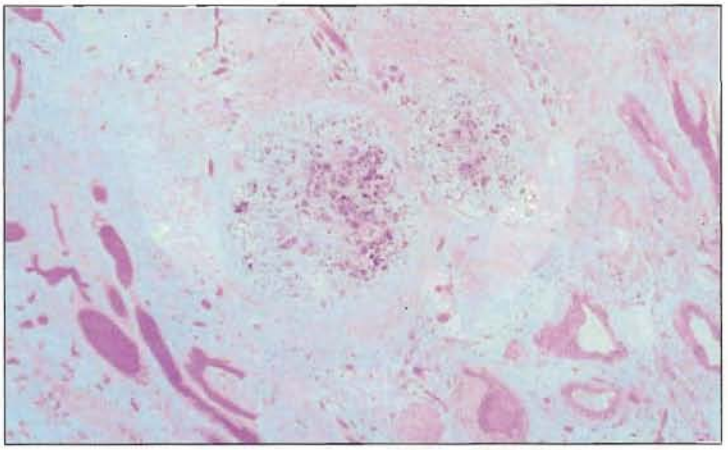

Figure 3—Photomicrograph of a section through the tip of the Meckel's diverticulum showing a 2.5-mm carcinoid tumor (hematoxylin and eosin stain, magnification x2), which was confirmed using chromogranin A staining (not pictured).

Most Meckel's diverticula are asymptomatic and found incidentally during surgery.[11] The male-to-female incidence ratio is approximately 2.4 to 1.[12] The lifetime risk of becoming symptomatic is about 4% to 6%, with most symptomatic cases occurring during childhood. The risk of complications generally decreases with increasing age.[11,13] Symptoms of Meckel's diverticula are commonly abdominal pain, nausea, vomiting, rectal bleeding, and abdominal distention.[14] When symptomatic, the most frequent findings are intestinal obstruction (36%), intussusception (13%), inflammation (12%), and hemorrhage (11%).[12] Heterotopic mucosa is found in up to 60% of Meckel's diverticula, of which more than 60% is gastric mucosa. Other types of heterotopic mucosa include pancreatic mucosa, colonic mucosa, endometrial mucosa, Brunner's glands, and hepatobiliary tissue.[3] In a 600-patient study by Yamaguchi and colleagues, neoplasm was also found in 3.2% of patients.[12]

Intussusception is the invagination of a segment of intestine (the intussusceptum) into an adjacent part of the gastrointestinal tract (the intussuscipiens). Intussusception in the pediatric patient is idiopathic about 90% of the time. With increasing age, the incidence of a pathologic lead point increases and is present in more than 50% of individuals older than 4 years of age. A study by Ong and Beasley showed that, in 27 of 56 patients (48%), the most common cause of intussusception was a Meckel's diverticulum.[15] Intussusception in adults is rare, with a mechanical process being the cause in 90% of cases.[16] Because most adult intussusceptions result from other conditions, they are often irreducible and require surgical intervention.[17]

Intussusception in the adolescent and adult populations can be difficult to diagnose. Patients' symptoms frequently differ from the classic symptoms of vomiting, acute abdominal pain, and lower gastrointestinal bleeding that occur in children. The history and physical examination of adults more often suggest an intermittent small bowel obstruction. Fewer than half of adults have a mass present, and peritoneal signs are uncommon.

Intussusception in the pediatric patient is idiopathic about 90% of the time.

Plain abdominal radiographs usually show signs of bowel obstruction with intussusception but are nonspecific regarding cause. Testing with oral Gastrografin may be useful at the time of symptoms and sometimes shows a characteristic beak-like point of obstruction. In many instances, however, the obstruction is only partial or intermittent, therefore limiting the usefulness of contrast studies. CT scanning has become the method of choice for diagnosing intussusception. The intussuscepted segment features a pathognomonic target lesion (Figure 1).[17] As the unrelieved obstruction progresses, bowel wall thickening and increasing intraluminal fluid produce the appearance of a mass at the 24-hour interval.[17] The CT

scan of the patient in this case report suggested perforated appendicitis with a multiloculated pelvic abscess. Because his symptoms had been present for some time, the inflamed, edematous bowel had the appearance of an abscess. Retrospectively, the pathognomonic target lesion can be identified on the CT scan (Figure 1).

Preoperative diagnosis of a Meckel's diverticulum can be a challenge for the physician. Abdominal pain is the most frequent symptom, followed by nausea, vomiting, and lower gastrointestinal bleeding.[12] Harkins and Ponka identified clinical features that help distinguish intussusception caused by a Meckel's diverticulum from other etiologies. Included in their findings were more frequent previous attacks, chronic pain, intense vomiting, a mass in the right lower quadrant, and less rectal bleeding than other lesions.[17] Past experience has shown that standard radiographic studies using contrast media are of little help in identifying a Meckel's diverticulum.[11] Radionucleotide scans can be used to identify ectopic gastric tissue that may be found in a diverticulum. Because this is an inconsistent finding, however, its usefulness is also limited.[11]

Carcinoid tumors arise most frequently from the gastrointestinal tract (73.5%) and bronchopulmonary system (25%). Most carcinoid tumors of the gastrointestinal tract occur in the small bowel (29%) and appendix (19%), followed by the rectum (12.5%).[18] A carcinoid tumor arising from a Meckel's diverticulum was first described in the literature in 1907.[19] Slightly more than 100 cases have since been documented, mostly in the form of case reports or in a review of the literature by Nies and colleagues.[8] The average age in their study was 56.8 years (range, 14–80 years), with 74% of patients older than 50 years and fewer than five patients younger than 20 years. The patients followed were predominantly asymptomatic (58 of 90; 64%, with the classic carcinoid syndrome occurring in only 8% [8 of 98] of patients). The average diameter of the tumors was 8.6 mm, with 72% of the tumors located in the tip. Significant findings included metastasis, seen at the time of diagnosis in 24% of patients.[8]

Only 1% to 2% of gastrointestinal carcinoid tumors are located in Meckel's diverticula.

Because carcinoid tumors in Meckel's diverticula are so rare, most are found incidentally on postmortem examination or at laparotomy for other conditions.[20] Only 1% to 2% of gastrointestinal carcinoid tumors are located in Meckel's diverticula.[8] They occur four times more often in men than in women, which has been attributed to the higher incidence of Meckel's diverticula and small intestinal carcinoid tumors in men.[21] These tumors resemble appendiceal carcinoid tumors in that they are usually small, singular, and asymptomatic. Because of their considerable metastatic potential, however, carcinoid tumors in Meckel's diverticula have been linked to jejunoileal carcinoid tumors.[19] The size of a carcinoid tumor is used as a determinant of metastatic potential by many clinicians.[22] Nies and colleagues found a metastatic rate of more than 50% in tumors larger than 10 mm.[8] Metastasis has been reported even with a primary tumor of 5 mm.[8] It has been emphasized by many that all extra-appendiceal carcinoid tumors should be considered malignant because of their relative aggressiveness.[8,22]

Because of the limited number of patients with carcinoid tumor of a Meckel's diverticulum that are available for follow-up evaluation, it is difficult to predict prognosis.[21] For adequate treatment, regional lymphadenectomy should be done, along with removal of a segment of ileum and the corresponding mesentery

for any carcinoid tumor of a Meckel's diverticulum larger than 5 mm.[8,22]

Conclusion

Our case report describes a rare occurrence of simultaneous pathologies. We hypothesize that the carcinoid tumor in our patient's Meckel's diverticulum was the lead point of the intussusception of the diverticulum into itself. The presence of ectopic gastric mucosa might also have been a factor. It is therefore likely that this intussuscepted Meckel's diverticulum functioned as the lead point in the intussusception of the ileum. This combination of occurrences, along with the rarity of finding a carcinoid tumor in a patient of this age, makes this case unique. ●●

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