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Rare presentation of pancreatitis secondary to intussusception of duodenal duplication cyst, a pediatric case report

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ABSTRACT

Duodenal duplication cysts are rare congenital malformations of the intestinal tract, for which there is limited literature in the pediatric population. The most common presentation in symptomatic patients is abdominal pain and pancreatitis. We present a case of a 14 year old female that presented with emesis, abdominal pain, weight loss, and admission biochemical profile concerning for acute pancreatitis in conjunction with severe hypochloremic, hypokalemic metabolic alkalosis. Further imaging was highly suggestive of duodeno-duodenal intussusception causing obstruction of the pancreatic duct. Patient was taken emergently to the operating room for exploration. Patient underwent laparoscopic assisted reduction of intussusception and resection of duodenal duplication cyst. Patient tolerated the surgery well, and was able to be discharged home in stable condition soon after. There have been no cases reported in the literature that describe pancreatitis secondary to intussusception of duodenal duplication cyst. When diagnosed early, these patients can be safely managed laparoscopically even in emergent settings.

1. Case report

A 14 year old African American female, with no significant past medical history, presented to the emergency department with several week history of non-bloody, non-bilious emesis and 10 pound weight loss. On physical exam, her abdomen was soft, non-distended, with moderate intermittent tenderness to palpation in the mid- and left epigastrum. At the time of admission, the patient was found to have profound electrolyte abnormalities with hypochloremic, hypokalemic metabolic alkalosis secondary to persistent emesis, as well as elevated pancreatic enzymes (Table 1). The patient’s electrolyte abnormalities were slow to respond to aggressive intravenous fluid resuscitation and electrolyte supplementation. To further work-up the source of the patient’s pancreatitis, an abdominal ultrasound was obtained, demonstrating concern for a possible ileocolic intussusception with a fluid-filled cystic lesion, acting as a potential lead point (Fig. 1). As these imaging findings were inconsistent with the patient’s initial presentation of pancreatitis, a CT of the abdomen/pelvis was obtained for further characterization. The scan demonstrated malrotation, with suggestion of duodeno-duodenal intussusception, with a cystic lesion acting as the lead point. Biliary ductal dilatation, as well as abnormal dilation of the pancreatic duct, was seen (Fig. 2). Given the grave concern for pancreatic duct obstruction with elevated pancreatic enzymes, and the risk for clinical deterioration secondary to pancreatic ductal obstruction as well as bowel wall ischemia and necrosis secondary to the intussusceptions, the patient and the mother were counseled to proceed with emergent surgical intervention despite gross electrolyte abnormalities. With parental consent, the patient was taken emergently to the operating room for exploration 3 h after initial surgical evaluation (24 h after admission to the hospital).

Diagnostic laparoscopy was performed. Duodenal intussusception, as suspected, was identified arising anterograde, from the second into the third portion of the duodenum, and reduced...
laparoscopically. It became obvious with reduction that a large duplication cyst encompassed the intussusception and a duodenal cyst resection was necessary. We laparoscopically mobilized the duodenum to identify the vena cava assuring there were no remaining Ladd’s bands prior to exteriorization. With the cecum positioned high and on the right, a wide base of mesentery available, the mobilization was all that was required to address the radiographically diagnosed malrotation. The umbilical incision was extended superiorly to allow delivery of the duplication cyst out through the umbilicus. A finger could be passed around the most proximal portion of the exteriorized bowel, confirming its distance away from the pancreatic head enabling complete cyst excision (Fig. 3). The proximal margin of the excised cyst, was close to the ampulla, however there was just enough distance to allow for resection and careful duodenoduodenostomy by end-to-end closure. The patient tolerated the procedure well and was discharged home on post-operative day six. Final pathology was consistent with duodenal duplication cyst (Fig. 4).

2. Discussion

Intestinal duplication cysts are extremely rare congenital malformations of the digestive tract, with an incidence of less than 1 per 100,000 live births. To be considered a duplication cyst, they must have an attachment to the native intestinal tract, a smooth muscle layer, and intestinal mucosal lining [2]. They are least likely to be located in the duodenum, with an incidence of 2–12% [1]. The most common presenting symptom is abdominal pain; the most common associated finding is pancreatitis. Intussusception of a duodenal duplication cyst is very rare, of which there have only been two other reports in the literature [3,4]. Other, more commonly described lead points causing duodenal intussusceptions, described in the literature include duodenal adenomas, lipomas, hamartomatous polyps, and hyperplasia [5]. The duodenum is not particularly susceptible to intussusception because the proximal duodenum is fixed around the head of the pancreas. The remainder is retroperitoneal, as it courses under the base of the mesentery, where the distal end is tethered by the ligament of Treitz as it emerges from the retroperitoneum. In this case, the intussusception was made possible by malrotation, resulting in a free intraperitoneal duodenum beyond the pancreas. This anatomy also allowed the duodenum to be easily brought out through the umbilicus.

Table 1

<table>
<thead>
<tr>
<th>Laboratory study</th>
<th>Value on day of admission</th>
<th>Reference range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium (mmol/L)</td>
<td>133 (L)</td>
<td>135–145</td>
</tr>
<tr>
<td>Potassium (mmol/L)</td>
<td>1.9 (L)</td>
<td>3.5–5.2</td>
</tr>
<tr>
<td>Chloride (mmol/L)</td>
<td>61 (L)</td>
<td>99–112</td>
</tr>
<tr>
<td>Carbon dioxide (mmol/L)</td>
<td>53 (H)</td>
<td>20–30</td>
</tr>
<tr>
<td>Anion gap (mmol/L)</td>
<td>32 (H)</td>
<td>7–14</td>
</tr>
<tr>
<td>BUN (mg/dL)</td>
<td>29 (H)</td>
<td>5–20</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
<td>0.9 (H)</td>
<td>0.35–0.84</td>
</tr>
<tr>
<td>Total bilirubin (mg/dL)</td>
<td>2.1 (H)</td>
<td>0–1.2</td>
</tr>
<tr>
<td>Direct bilirubin (mg/dL)</td>
<td>0</td>
<td>0–0.4</td>
</tr>
<tr>
<td>AST (unit/L)</td>
<td>101 (H)</td>
<td>12–50</td>
</tr>
<tr>
<td>ALT (unit/L)</td>
<td>65 (H)</td>
<td>5–50</td>
</tr>
<tr>
<td>Alk phos (unit/L)</td>
<td>126</td>
<td>70–230</td>
</tr>
<tr>
<td>LDH (unit/L)</td>
<td>688 (H)</td>
<td>370–645</td>
</tr>
<tr>
<td>Amylase (unit/L)</td>
<td>200 (H)</td>
<td>3–110</td>
</tr>
<tr>
<td>Lipase (unit/L)</td>
<td>1040 (H)</td>
<td>23–300</td>
</tr>
</tbody>
</table>

Fig. 1. Classic target sign appearance of intussusception noted in right upper quadrant, measuring 5.3 × 4.4 cm, initially concerning for possible ileocolic intussusception.

Fig. 2. Axial cut of abdominal CT demonstrating abnormal dilation of the pancreatic duct, as well as duodenal intussusception.

Fig. 3. Excised duodenal duplication cyst (3 × 2 × 2.5 cm) through an opening of the dilated proximal duodenum being held by clamp. Forceps are within the lumen of distal duodenum. The ampulla was identified at the resection margin.
To our knowledge this is the only case of duodenal duplication cysts presenting with pancreatitis, secondary to duodeno-duodenal intussusceptions, in both the pediatric and the adult populations. This case also demonstrates how the obstructive type pattern can create significant electrolyte abnormalities that could have serious negative consequences if immediate intervention is not taken.

Optimal treatment of symptomatic duodenal duplication cysts is complete surgical resection. The relationship of the duplication cyst with the pancreatic or biliary ducts can affect the type of procedure that is performed, including: complete surgical removal, partial resection, marsupialization of the cyst, and even pancreaticoduodenectomy [1,6]. Successful laparoscopic, as well as endoscopic, management has been described in the pediatric population [7,8]. Our case demonstrates that the minimally invasive method of laparoscopically-assisted resection, even in emergent settings, can be performed safely in the pediatric population with successful outcomes.

3. Conclusion

Presentation of pancreatitis secondary to intussusception of duodenal duplication cyst is very rare, for which there have been no other reports in the literature. With swift diagnosis and intervention, these patients can be safely managed laparoscopically, with favorable outcomes even in emergent settings.

References