Gastric outlet obstruction caused by an ectopic pancreas in a neonate: A case report

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Gastric outlet obstruction caused by an ectopic pancreas in a neonate: A case report

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\textbf{A B S T R A C T}

We herein report a neonate who presented with non-bilious vomiting at one day of age caused by a prepyloric ectopic pancreas. Ultrasonography clearly detected the presence of a submucosal mass preoperatively, which was treated with local gastric resection. Only 9 neonates with a symptomatic pyloric or prepyloric ectopic pancreas have been previously reported in the literature. Therefore, we reviewed and discussed the clinical features of neonates with this type of ectopic pancreas.

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Ectopic pancreas is defined as pancreatic tissue that lacks anatomic and vascular continuity with the main body of the pancreas [1]. It is commonly asymptomatic and incidentally found in the stomach, duodenum or upper part of the jejunum during operations and at autopsy [2]. Symptomatic ectopic pancreas in neonates is extremely rare. We herein report a one-day-old female neonate who presented with non-bilious vomiting caused by an obstruction secondary to an ectopic pancreas located at the prepyloric antrum.

\section*{1. Case report}

A 3125 g female baby, one of dichorionic diamniotic twins, was born at 37 weeks of gestation by cesarean section, indication for which was a previous cesarean delivery. Her prenatal surveys had revealed normal amounts of amniotic fluid. Her Apgar scores were 8 and 9 at 1 and 5 min after birth, respectively. Her oral intake was started soon after birth as usual, however, she presented with non-bilious vomiting 4–6 times per day on the first day after birth. She had been conservatively observed in the newborn nursery until six days after birth. A physical examination at that time revealed a soft abdomen and normal bowel sounds without a palpable mass. The patient’s body weight was 2768 g, which showed an 11.4% decrease compared to her birth body weight. Her blood tests were unremarkable except for mild hyperbilirubinemia. An abdominal X-ray demonstrated a gas in the large intestine predominantly and did not indicate a complete bowel obstruction. Abdominal ultrasonography (US) was then performed, which revealed a submucosal solid mass (measuring approximately 7 mm in diameter) at the anterior wall of the prepyloric gastric antrum (Fig. 1). The width of the pyloric wall and length of the pyloric channel were within normal limits, which excluded hypertrophic pyloric stenosis (HPS). An upper gastrointestinal (UGI) series demonstrated a gas in the large intestine predominantly and did not indicate a complete bowel obstruction. Abdominal ultrasonography (US) was then performed, which revealed a submucosal solid mass (measuring approximately 7 mm in diameter) at the anterior wall of the prepyloric gastric antrum (Fig. 1). The width of the pyloric wall and length of the pyloric channel were within normal limits, which excluded hypertrophic pyloric stenosis (HPS). An upper gastrointestinal (UGI) series demonstrated a narrow pylorus and a round-shaped defect of the contrast medium at the prepylorus in the prone position, potentially due to the above mentioned mass (Fig. 2). Thus, a jejunal tube was placed for enteral feeding. Contrast-enhanced abdominal computed tomography (CT) was performed to confirm the characteristics of the mass and it localized the slightly enhanced mass at the same site indicated on US. The preoperative diagnosis was a submucosal mass at the prepyloric gastric antrum, which included the differential diagnoses of ectopic pancreas and gastric duplication. At 16 days of age, the infant underwent exploratory laparotomy via an upper half circumumbilical incision. The mass was palpated at the anterior
wall of the antrum and excised with a full thickness of the local gastric wall (Fig. 3). The defect of the antral wall was closed in a single layer with interrupted sutures. It had a central umbilication on the mucosal surface of the mass. The postoperative course was uneventful except for slow recovery of gastric emptying. She was discharged on the 24th postoperative day and has gained normal growth and development during the follow-up period of 18 months. The excised mass specimen consisted of a 10 × 10 × 8 mm, solid, milk-white colored tissue. A histologic examination revealed aberrant submucosal exocrine pancreatic tissue that had acini, ducts, and islets cells, which was compatible with an ectopic pancreas (Fig. 4).

2. Discussion

Ectopic pancreas occurs in 1%–2% of autopsies and in 1:500 laparotomies with a male to female ratio of 3:1 [1]. Ninety percent of all cases are found in the stomach, duodenum, and jejunum, although it can be found throughout the gastrointestinal tract and in other intra-abdominal organs [2]. The majority of patients with ectopic pancreas are clinically asymptomatic. Its clinical manifestation may include some degree of obstruction resulting from the enlarged nodule or complications such as inflammation, ulceration, tumor, or intussusception [3].
To the best of our knowledge, only 9 neonatal cases with symptomatic ectopic pancreas located at the pylorus or prepyloric antrum have been previously reported in the literature [4–12]. Table 1 shows the demographics and clinical outcomes of these cases, including ours. The onset of the symptoms ranged from one day to 21 days of age. Most of the initial symptoms were vomiting. Gastrointestinal bleeding was noted in four cases (PT#4, 6, 9, 10), leading to additional imaging studies for the differential diagnosis because it may visualize the ectopic pancreas as a pyloric or prepyloric mass, such as in our case. The UGI series may visualize the submucosal mass with central umbilication, but may only show a narrowed pylorus and may not be consistent with an ectopic pancreas. Endoscopy would be able to visualize the submucosal mass with a central umbilication as a typical feature of the ectopic pancreas, however, it requires experienced pediatric gastroenterologists or surgeons who are familiar with this procedure under general anesthesia for neonates. CT with contrast enhancement may detect the ectopic pancreas as a mass and may rule out other abnormalities, but its merit should be weighed against its risk of radiation exposure. If abdominal US clearly detects the submucosal mass around the gastric outlet, then that would be adequate for diagnosis. A correct diagnosis and precise localization of the ectopic pancreas would avoid unnecessary operations or overtreatment, thereby leading to its local excision with a minimal incisional approach, such as a circumumbilical incision as was performed in our case or a laparoscopic approach.

References


Table 1
Neonatal cases with symptomatic ectopic pancreas located at the pylorus or prepyloric antrum.

<table>
<thead>
<tr>
<th>Patient no</th>
<th>Author</th>
<th>Year</th>
<th>Sex</th>
<th>Onset</th>
<th>Symptom(s)</th>
<th>Preoperative imaging(s)</th>
<th>Operation</th>
<th>Age at operation</th>
<th>Time of final diagnosis</th>
<th>Outcome</th>
<th>Size</th>
</tr>
</thead>
<tbody>
<tr>
<td>#1</td>
<td>Kernohan</td>
<td>1956</td>
<td>M</td>
<td>10 days</td>
<td>Non-bilious vomiting</td>
<td>HPS</td>
<td>UGI series, Ramstedt’s operation</td>
<td>20 days</td>
<td>At autopsy</td>
<td>Died</td>
<td>NA</td>
</tr>
<tr>
<td>#2</td>
<td>Matsumoto</td>
<td>1974</td>
<td>F</td>
<td>5 days</td>
<td>Non-bilious vomiting and melena</td>
<td>HPS</td>
<td>Physical exam</td>
<td>7 days 6 months</td>
<td>After 2nd operation</td>
<td>Alive</td>
<td>NA</td>
</tr>
<tr>
<td>#3</td>
<td>Ishihara</td>
<td>1980</td>
<td>M</td>
<td>20 days</td>
<td>Non-bilious vomiting</td>
<td>HPS</td>
<td>Endoscopy, US, Subserosal excision</td>
<td>8 days 1st operation</td>
<td>Alive 3 mm</td>
<td>Alive</td>
<td>NA</td>
</tr>
<tr>
<td>#4</td>
<td>Visentin</td>
<td>1991</td>
<td>M</td>
<td>18 days</td>
<td>Non-bilious vomiting</td>
<td>Hypertrophic pyloric mass</td>
<td>US (1) Biopsy, gastrojejunostomy</td>
<td>18 days 2 yo</td>
<td>Biopsy</td>
<td>Alive</td>
<td>NA</td>
</tr>
<tr>
<td>#5</td>
<td>Ueno</td>
<td>1993</td>
<td>F</td>
<td>10 days</td>
<td>Hematemesis and melena</td>
<td>Submucosal tumor</td>
<td>Endoscopy, US</td>
<td>6 months</td>
<td>After operation</td>
<td>Alive</td>
<td>3-4 mm</td>
</tr>
<tr>
<td>#6</td>
<td>Hayes-Jordan</td>
<td>1998</td>
<td>M</td>
<td>2 days</td>
<td>Non-bilious vomiting</td>
<td>Submucosal tumor</td>
<td>Subserosal excision</td>
<td>3 days</td>
<td>After operation</td>
<td>Alive</td>
<td>4-5 mm</td>
</tr>
<tr>
<td>#7</td>
<td>Ozcan</td>
<td>2002</td>
<td>M</td>
<td>7 days</td>
<td>Non-bilious vomiting</td>
<td>Submucosal tumor</td>
<td>Partial gastrectomy</td>
<td>40 days</td>
<td>Alive NA</td>
<td>Alive</td>
<td>NA</td>
</tr>
<tr>
<td>#8</td>
<td>Fragoso</td>
<td>2004</td>
<td>M</td>
<td>21 days</td>
<td>Non-bilious vomiting</td>
<td>Ectopic pancreas</td>
<td>Endoscopy, US</td>
<td>18 yo</td>
<td>Alive NA</td>
<td>Alive</td>
<td>NA</td>
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<tr>
<td>#9</td>
<td>Surov</td>
<td>2015</td>
<td>F</td>
<td>12 days</td>
<td>Hematemesis and melena</td>
<td>Arteriovenous malformation</td>
<td>US, CT</td>
<td>Resection of tumor with local gastric wall</td>
<td>16 days</td>
<td>After operation 10 mm</td>
<td>Alive</td>
</tr>
<tr>
<td>#10</td>
<td>Our case</td>
<td>2015</td>
<td>F</td>
<td>1 day</td>
<td>Non-bilious vomiting</td>
<td>Ectopic pancreas, gastric duplication</td>
<td>US, UGI series, CT</td>
<td>Alive 4-5 mm</td>
<td>Alive 10 mm</td>
<td>Alive</td>
<td>NA</td>
</tr>
</tbody>
</table>

To the best of our knowledge, only 9 neonatal cases with symptomatic ectopic pancreas located at the pylorus or prepyloric antrum have been previously reported in the literature [4–12]. Table 1 shows the demographics and clinical outcomes of these cases, including ours. The onset of the symptoms ranged from one day to 21 days of age. Most of the initial symptoms were vomiting. Gastrointestinal bleeding was noted in three cases (PT#2, 5, 9). The clinical courses of the first four cases (PT#1–4) were not straightforward; PT#1 had Ramstedt’s operation without treating the lesion and subsequently died of gastrointestinal bleeding. PT#2 and #3 had two operations because the primary operations only revealed the pyloric mass, which necessitated antrectomy later in secondary operations. PT#4 underwent gastrojejunostomy and a biopsy of the prepyloric lesion in the primary operation, revealing the ectopic pancreas. The physicians decided to observe the patient without removing the lesion and had the anastomosis detached, thereby regaining normal intestinal continuity in the secondary operation.

A submucosal tumor at the prepyloric lesion was detected by endoscopy in three cases (Pt#5, 6, 8), and a central umbilication on the mucosa was also noted in Pt#5. An UGI series was performed in six cases (Pt#1, 3, 5, 6, 7, 10); a prepyloric mass was demonstrated in three cases (Pt#5: prepyloric mass with a central umbilication, #6 and #10: antral mass), and partial or complete pyloric obstruction was the only finding in the remaining cases. When the imaging study was limited to the UGI series, a correct diagnosis was hard to obtain preoperatively (Pt#1, 3, 7). Abdominal US was able to detect the prepyloric mass in four cases (Pt#4, 6, 9, 10), leading to additional imaging studies on the suspicion of pathologies other than HPS in three cases (Pt#6, 9, 10).

HPS is most often encountered as a cause of non-bilious vomiting in early infancy, and a physical examination and abdominal US, revealing a palpable enlarged pylorus and hypertrophied pyloric muscle with elongated pyloric channel, respectively, are common diagnostic methods for HPS [13]. If these examinations do not confirm HPS, then the differential diagnoses include various conditions such as pylorospasm, gastroesophageal reflux, gastric volvulus, antral web, preampullary duodenal stenosis, duplication cyst, and ectopic pancreas [13]. Abdominal US remains important for the differential diagnosis because it may visualize the ectopic pancreas as a pyloric or prepyloric mass, such as in our case. The UGI series may visualize the submucosal mass with central umbilication, but may only show a narrowed pylorus and may not be consistent with an ectopic pancreas. Endoscopy would be able to visualize the submucosal mass with a central umbilication as a typical feature of the ectopic pancreas, however, it requires experienced pediatric gastroenterologists or surgeons who are familiar with this procedure under general anesthesia for neonates. CT with contrast enhancement may detect the ectopic pancreas as a mass and may rule out other abnormalities, but its merit should be weighed against its risk of radiation exposure. If abdominal US clearly detects the submucosal mass around the gastric outlet, then that would be adequate for diagnosis. A correct diagnosis and precise localization of the ectopic pancreas would avoid unnecessary operations or overtreatment, thereby leading to its local excision with a minimal incisional approach, such as a circumumbilical incision as was performed in our case or a laparoscopic approach.

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