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Case Report

Duodenal Stenosis: A Case Report

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Abstract:

Background: Congenital duodenal stenosis in pediatric patients was often underreported due to its non-conspicuous signs and symptoms. Diagnosing duodenal stenosis is often challenging as this disease causes partial intestinal obstruction and thus presents with more indolent and atypical clinical manifestations. This case report aims to describe the atypical case of pediatric duodenal stenosis which presented with recurrent vomiting and poor weight gain as well as highlight some of the diagnostic challenges.

Case: A 7-month-old girl was admitted to the emergency room with chief complaint of recurrent vomiting in the last 2 days prior to hospital admission. Patient had a history of recurrent bilious vomiting at the age of 3 days old with a frequency of 3-4 times a day and were admitted to the hospital for 2 weeks. Parents also reported of poor weight gain in the last 3 months. Abdominal X-Ray series showed dilatation of the small intestines immediately after pylorus and stack of coins sign. Esophageal endoscopic evaluation showed signs of severe GERD with a pyloric gap as well as a suspicion of a duodenal web

Discussion: Congenital obstruction at the duodenum may occurs due to intrinsic or extrinsic etiology. Failure of duodenal re-canalization during the 8-10th week of embryological development is thought to be the main cause of intrinsic duodenal obstruction (atresia, stenosis or duodenal web). The appearance of clinical manifestation of duodenal stenosis depends on the degree of stenosis itself.

Conclusion: Congenital duodenal stenosis may present with atypical presentations in neonates which requires clinicians to be fully aware of this diagnosis to ensure timely therapy. The main management of duodenal stenosis is surgery, however fluid administration, decompressing as well as other supportive treatment are equally crucial to ensure better outcome for the patient.

Keywords: duodenal stenosis, pediatric, poor weight gain, vomiting

Introduction

Congenital duodenal stenosis in pediatric patients was often underreported due to its non-conspicuous signs and symptoms. Diagnosing duodenal stenosis is often

challenging as this disease causes partial intestinal obstruction and thus presents with more indolent and atypical clinical manifestations. Poor weight gain is one of the most common symptoms in children with duodenal stenosis. However, this clinical manifestation often undetectable by the parents due to inappropriate feeding practice or feeding the children with concentrated infant formula.[1] In comparison, in the case of complete small bowel obstruction such as duodenal atresia or volvulus, most patients present with acute and severe signs and symptoms of obstruction such as profuse bilious vomiting.

The etiology of duodenal obstruction can be classified as intrinsic or extrinsic based on the source of blockage.[2] Intrinsic duodenal obstruction refers to duodenal atresia, duodenal stenosis and duodenal webs.[3] The mechanism behind this is thought to be due to failure of the duodenal lumen to recanalize during fetal development, which occurs at roughly 8-10 weeks in utero.[3] On the other hand, extrinsic duodenal obstruction may occur due to malrotation, midgut volvulus, annular pancreas or any other external organs that compress the lumen of duodenum. [4]

For clinicians, initial evaluation of children with suspected bowel obstruction should focus on differentiating whether the cause of that obstruction is an emergency or not, to avoid further devastating consequences. Therefore, this case report aims to describe the atypical case of pediatric duodenal stenosis which presented with recurrent vomiting and poor weight gain as well as highlight some of the diagnostic challenges.

Case

A 7-month-old girl was admitted to the emergency room with chief complaint of recurrent vomiting in the last 2 days prior to hospital admission. Complaint was also accompanied by fever that started 1 day prior to admission. Vomiting occurred with a frequency of 4-5 times per day, which consisted of milk, white in color with no clumping. Parents reported that the patient vomited about as much as 1 aqua cup during the first episode of vomiting and then half aqua cup at the second episode of vomiting.

According to the patient's mother, fever was not immediately high, although it was not properly measured, and continued throughout the day with no temperature fluctuation. She also noticed that her child had watery stool 1 day prior to admission with a frequency of 4 times a day and as much as half aqua cup for each bowel movement. Stool was characterized as brown in color with foul odor which was different than usual, but without any mucous or blood. According to the patient's mother, the patient could still drink but the patient looked very thirsty. In the

emergency room after the patient was given fluids through IV access, the patient didn't seem thirsty anymore.

Patient had a history of recurrent bilious vomiting at the age of 3 days old with a frequency of 3-4 times a day and were admitted to the hospital for 2 weeks. Parents also reported of poor weight gain in the last 3 months.

On physical examination, patient was fully conscious but looked moderately ill. Vital signs were normal. Patients had malnutrition with body weight of 6 kg and body length of 66 cm (z score for weight-for-length <-2 SD). Nothing remarkable during inspection of the abdomen, however pain was felt during palpation with no organomegaly. No shifting dullness was observed during abdominal percussion. Bowel movement was 6 times per minute.

Upon laboratory evaluation, hypochromic microcytic anemia, leukocytosis and thrombocytosis were found (**Table 1**).

Table 1. Laboratory results from the patient

Parameters (Unit)	Value	Reference Value
Hemoglobin (g/dL)	10.6	10.5-14.0
Hematocrit (%)	33.3	32-42
Erythrocyte (10^6/µL)	4.58	3.95-5.26
MCV (fL)	72.7	72-88
MCH (pg)	23.1	24-30
MCHC (%)	31.8	32-36
Thrombocyte (10^3/mm^3)	609	150-400
Leucocyte (10^3/mm^3)	18.85	6-14
Neutrophil (%)	77.7	25-60
Monocyte (%)	6.8	2-8
Lymphocyte (%)	9.1	20-40
Eosinophil (%)	6.2	1-3
Basophil (%)	0.2	0-1
ESR	25	0-20
Calcium ion (mmol/L)	1.02	1.01-1.31
Phosphate (mg/dL)	6.3	2.5-7.0
Magnesium (mg/dL)	2.03	1.70-2.55

Two days after being treated, complaints of watery stool improved but the patient still experienced repeated profuse vomiting 3-4 times a day. Abdominal X-Ray with contrast was performed (**Figure 1**). Patient was then consulted to pediatric surgeon. Upon esophageal endoscopic evaluation, severe GERD with a pyloric gap was found

as well as a suspicion of a duodenal web (**Figure 2**). The patient was then planned for laparotomy correction of duodeno-duodenostomy. Patient was then treated with cefotaxime 150 mg three times a day, paracetamol 75 mg four times a day and intravenous fluid.

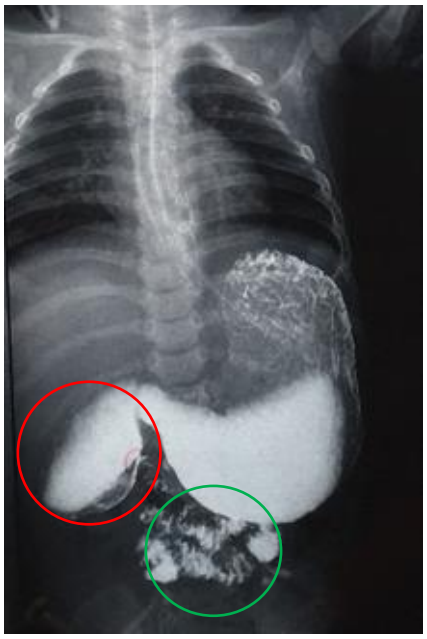

Figure 1. Abdominal X-Ray series showed dilatation of the small intestines immediately after pylorus (red circle) and stack of coins sign (valvulae conniventes) distal to the dilatation part (green circle).

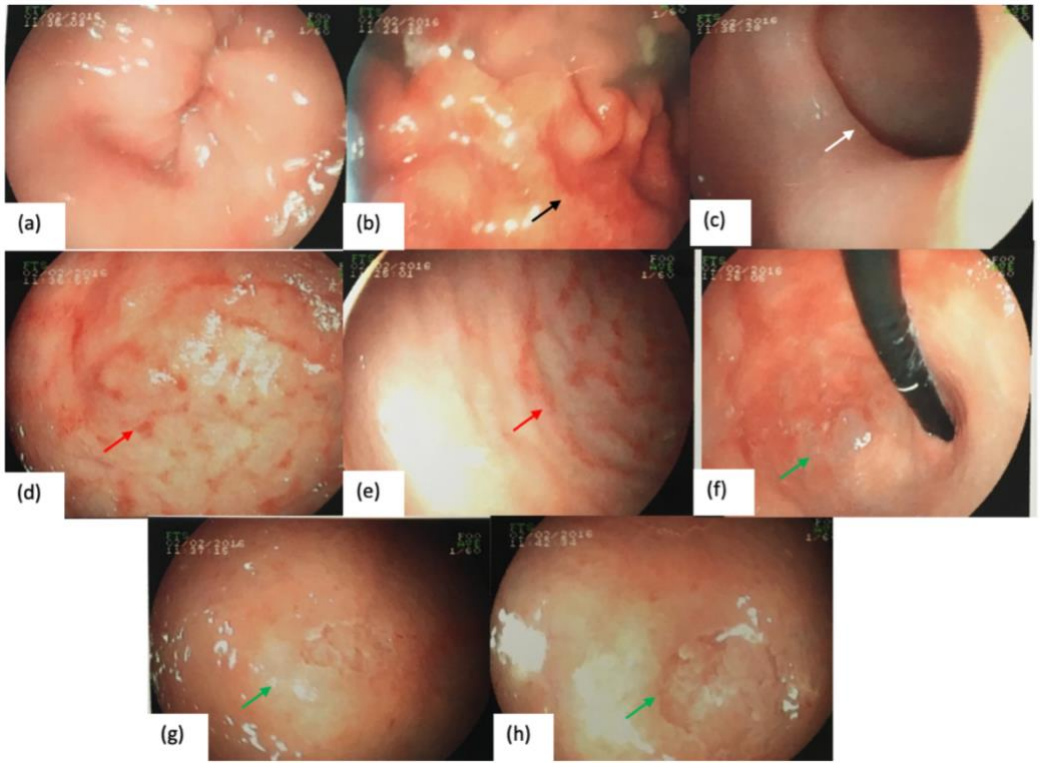

Figure 2. (a) Esophageal mucosa was normal. **(b)** Mucosa of the gastric corpus, cardiac and fundus showed remarkable hyperemia, erosion and gastropathy (black arrow). **(c)** Wide opening of the pylorus (white arrow). **(d), (e)** Mucosa of the duodenum was hyperemic with extensive erosion and fragile (red arrows). **(f), (g), (h)** Lumen of the duodenum was blocked by mucosal fold which prevented the scope to pass through (green arrows).

Discussion

Duodenal stenosis, along with duodenal atresia, is a congenital intestinal obstruction that leads to bilious or non-bilious vomiting within the first 24 to 72 hours of neonatal life, following the first oral feeding.[1,5] Duodenal stenosis and duodenal atresia is the most common cause of intrinsic congenital duodenal obstruction which occurs 1 in 5000 to 10,000 live births.[5]

Congenital obstruction at the duodenum may occurs due to intrinsic or extrinsic etiology. Intrinsic duodenal obstruction can be caused by duodenal atresia, duodenal stenosis, and duodenal webs. On the other hand, extrinsic duodenal obstruction can be caused by malrotation with Ladd's bands or by the presence of the preduodenal portal vein.[1,6]

Failure of duodenal re-canalization during the 8-10th week of embryological development is the main cause of duodenal atresia. It usually occurs at the distal of the ampulla of Vater, in the second portion of the duodenum.[5] Duodenal atresia

occurs when the duodenum is not completely formed which results in a complete obstruction of the duodenal lumen. In duodenal stenosis, the lumen is narrowing, resulting in an incomplete obstruction of the duodenum lumen. These anomalies are frequent in newborns and patients with chromosomal abnormalities, such as Down syndrome.[7] A duodenal web is a more rare cause of duodenal obstruction, in which the duodenal lumen tends to have a windsock deformity.[5,6] In this case, there was no evidence of an extrinsic obstruction.

Manifestation of duodenal stenosis appears within 24 to 72 hours after birth, in which the age of presentation depends on the degree of stenosis itself. The presentation may appear later compared to duodenal atresia due to more distal blockage, hence abdominal distension and vomiting were the most common features.[1] Vomiting occurs due to an obstruction in the upper digestive tract, hence the breast milk or amniotic fluid that pass through the stomach cannot proceed to the duodenum, thus resulting in vomiting a few hours after birth.[6] Clinical presentation may vary from repeated vomiting, gastric distension, failure to thrive in infancy, gastroesophageal reflux and peptic ulcer which depends on the age of the patients. Poor weight gain, signs of dehydration, minimal or no stool may also present.[1,7,8] This patient came to our hospital due to recurrent vomiting for two days, 4-5 times a day, with white in color without any lumpy texture. However, this patient also had a history of being treated for two weeks in the hospital due to recurrent vomiting within the first 72 hours of life, with bilious vomitus as frequent as 3-4 times. No other symptoms or abnormalities were recorded at birth. The presence of bilious emesis indicates duodenal atresia or stenosis. Obstruction distal to the ampulla of Vater results in a greenish color of bile mixed with vomit that comes out. The symptoms that appear depend on the location of the obstruction itself. Proximal obstruction of the intestinal tract may present as frequent bilious vomiting with large volume. Meanwhile, the distal obstruction is indicated by moderate abdominal distention with progressive vomiting. Patients with duodenal atresia or stenosis may develop dehydration when vomiting persists. It is also necessary to look for signs of dehydration such as sunken fontanels, reduced or even absent tears, dry oral mucosa, slowed skin turgor, and if the baby is looking weak and apathetic.[6] Our patient showed dehydration symptoms, in which she looked very thirsty when she first came. The symptom improved after adequate rehydration was given. The patient's body weight increased up to 3 months old, yet decreased afterward. Patient also suffered from malnutrition at the time of admission. Although there was some notable weight loss, this might also be related with previous hospitalization episode due to septic shock, meningitis, history of seizure, and loss of consciousness. Our patient also showed delayed in developmental milestones which needed further evaluation.

On inspection, abdominal distension may be seen, indicating distal obstruction. On palpation examination, the stomach in the epigastric area might feel enlarged.[6] Our patient showed flat abdomen on inspection, which might be due to previously installed nasogastric tube, resulting in decompression.

Laboratory findings may show hemoconcentration suggesting dehydration. Hypokalemia or hypochloremic metabolic alkalosis can also be found due to repetitive vomiting.[1,6] However, in this case, only leukocyte, neutrophil, lymphocytes, and erythrocyte sedimentation rate increased. Leukocytosis may be related with fever and diarrhea that presented 1 day before hospital admission in this patient. Other findings were within normal limits.

Duodenal stenosis may present with polyhydramnios or dilated loops of bowel on fetal sonography during prenatal period. Extrinsic duodenal obstruction such as annular pancreas, duplication cyst, and preduodenal portal vein as well as pyloric stenosis need to be ruled out in postnatal ultrasound.[1] None of these findings were showed in our cases.

Plain abdominal radiograph has been used as the first step for duodenal stenosis evaluation. The double bubble sign with the absence of distal intestinal gas indicates duodenal stenosis or atresia.[1,6,8] The first bubble indicates the presence of fluid filling in the gastric area. The second bubble indicates the distended post-pyloric/proximal duodenum which is located before the site of atresia or stenosis. No visible air in the rest of the small intestine or large intestine suggests duodenal atresia (complete obstruction), meanwhile, an uneven distribution of air in the distal part of the obstruction suggests the possibility of stenosis or volvulus (partial obstruction). Radiological examination using contrast is used to rule out the possibility of malrotation.[6] In this case, the patient's abdominal x-ray revealed a dilatation of the intestinal area after the pylorus and valvula conniventes (a stack of coins after the area is dilated). In addition, the results of esophagogastroduodenoscopy showed severe gastroesophageal reflux, gastropathy, pyloric gap, and suspected duodenal web. A duodenal web is a rare cause of duodenal obstruction, which tends to cause a windsock deformity of the duodenal lumen.[5] Hence, it is concluded that the main symptoms of our patient were caused by an intestinal obstruction at the level of the duodenum due to duodenal stenosis.

The management of duodenal stenosis involves several aspects. Fluid administration needs to be given according to the degree of dehydration. Decompression is done by inserting a nasogastric or orogastric tube.[6,8] Nutrition can be given parenterally according to the needs of the patient pre- and post-surgery. Gastric residual is usually monitored after the surgery. Electrolytes should be monitored, especially when

dehydration occurs. The optimal temperature also needs to be maintained. The definitive treatment of duodenal atresia or stenosis is surgery to resect the obstructed part and connect the remaining part of the digestive tract with an anastomosis.[6] The recommended surgical procedure is the duodeno-duodenostomy in which the proximal and distal sections of the duodenum are connected after resection of the obstruction site[6-8], which was performed in our patient. In this case, the fluid administration was immediately given when the signs of dehydration were found. The decompression had also been carried out by inserting a nasogastric tube. Antibiotics and antipyretics were also given.

Conclusion

Congenital duodenal stenosis may present with atypical presentations in neonates which requires clinicians to be fully aware of this diagnosis to ensure timely therapy. Diagnosing duodenal stenosis is often challenging as this disease causes partial intestinal obstruction and thus presents with more indolent and atypical clinical manifestations. Plain abdominal radiograph as well as series abdominal X-Ray with contrast are beneficial to confirm the diagnosis of duodenal stenosis as well as to exclude other differential diagnosis particularly those from extrinsic source. The main management of duodenal stenosis is surgery, however fluid administration, decompressing as well as other supportive treatment are equally crucial to ensure better outcome for the patient.

Conflict of Interest

None declared.

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