CASE REPORT

Delayed Diagnosis of Duodenal Atresia in an 11 Year Old

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Abstract

Duodenal atresia is a condition typically diagnosed in the neonatal period. Here, we discuss an 11-year-old patient with a new diagnosis of duodenal atresia, discovered during a lysis of adhesions. The patient had a history of malrotation and was status-post Ladd's procedure, but had continued to experience bilious emesis and symptoms of intestinal obstruction since 1 month of age, resulting in lifetime total parenteral nutrition (TPN) dependence. She was subsequently diagnosed with microcolon megacystic hypoperistalsis syndrome (MMHS) and underwent loop jejunostomy creation that proved unsuccessful in relieving her symptoms. Duodenal atresia was recognized and repaired intraoperatively during a planned loop jejunostomy revision.

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Introduction

Duodenal atresia is found in 1 out of every 5,000-10,000 live births, and is more common in males than females. Approximately 25% of patients with duodenal atresia also have Down syndrome.[1] It typically presents shortly after birth with bilious vomiting, and can be diagnosed on abdominal x-ray via a characteristic double-bubble sign. Treatment consists of nasogastric tube decompression of the stomach, fluid resuscitation, and duodenoduodenostomy. It is thought to be the result of intrauterine vascular insult, or failure of recanalization during intestinal development; as such, pregnancies can often exhibit polyhydramnios.[1] It should be considered in the differential for bilious emesis in the newborn.

As in the case of any complicated or delayed diagnosis, an incompletely characterized clinical picture has the potential to obscure symptomatology, and cloud medical decision making. Patients can be subject to debilitating disease sequelae or exposed to the risks of additional procedures and prolonged treatments. When considering duodenal atresia specifically, the clinical course could include fewer surgical interventions or a shorter timeframe of TPN dependence if identified and treated in a timely manner. Undoubtedly, patients with complex medical histories present challenges when considering the workup of a differential diagnosis. It is logical that as a patient ages, congenital malformations may be less likely to be considered as the source of persistent or refractory symptoms. For this reason, it is important to revisit these inborn errors as potential sources of dysfunction if they have not been formally or adequately ruled out in the diagnostic process.

Case Report

Our patient was an 11-year-old female with complete TPN dependence since birth secondary to frequent bilious emesis and longstanding obstructive symptoms. She had a diagnosis of MMHS in the setting of a history of a Ladd's procedure and gastrostomy tube (g-tube) placement at 1 month old. Despite these interventions, she remained unable to tolerate enteral feeds, exhibited recurrent episodes of emesis with abdominal pain, and required frequent fecal disimpactions. Small bowel follow through was attempted in February 2018 without progression of contrast beyond the duodenum at 5.5 hours, as evidenced in Figure 1.

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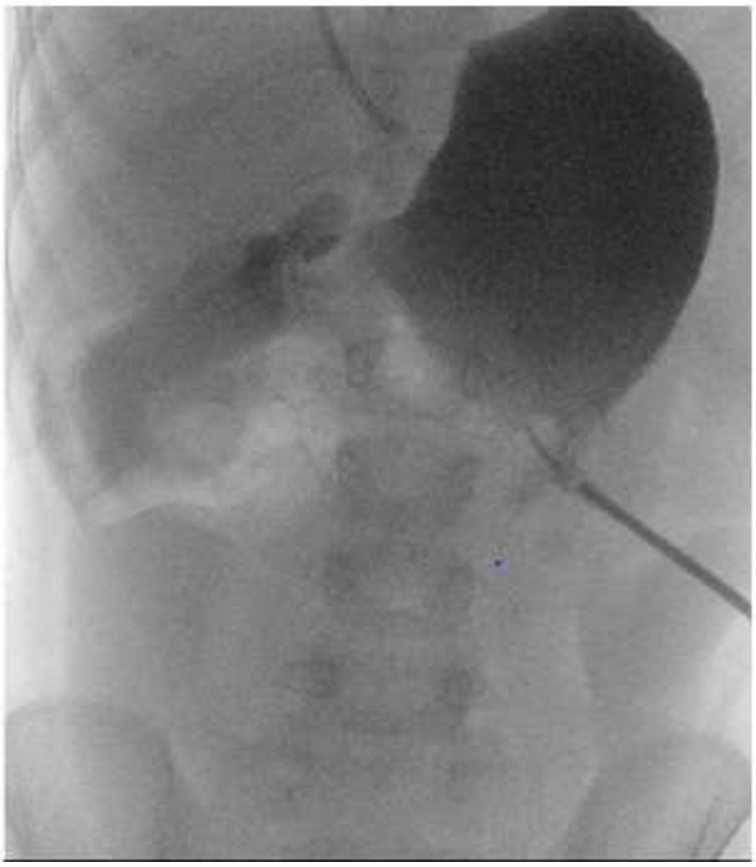

Figure 1: Small bowel follow through showing contrast to the level of the duodenum 5.5 hours into the study

At 10 years old she transferred care to our facility and underwent loop jejunostomy creation. She continued to have high-volume bilious output from her g-tube, and comparatively low-volume clear output from her loop jejunostomy. She also complained of distension, emesis, constipation, and difficulty urinating. Because of her persistent symptomatology, a loop jejunostomy revision was scheduled.

Extensive lysis of adhesions was performed. When the duodenum was isolated, an atretic band appeared to be present between portions 2 and 3 of the duodenum. A foley catheter was inserted into the gastrostomy and was unable to be passed across the atretic band. A duodeno-duodenostomy was then performed, and patency from portion 2 to portion 3 of the duodenum was confirmed by passing the foley catheter with the balloon inflated across the new anastomosis. We elected to proceed with the original plan of loop jejunostomy revision for obstructive relief, with a future goal of jejunostomy takedown pending symptomatic improvement.

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Discussion

Our patient is an 11-year-old girl without Down syndrome whose medical history of malrotation status-post Ladd's procedure complicated her persistent symptomatology. Her bilious emesis persisted after her initial loop jejunostomy because her ampulla of Vater was proximal to the atretic duodenal band. The loop jejunostomy had scant, non-bilious production because it was distal to the atretic segment of duodenum. Thus, the initial loop jejunostomy did not relieve her symptoms; the true cause of her intestinal obstruction and bilious emesis (duodenal atresia) was only discovered after extensive lysis of adhesions.

Interestingly, our patient did not have the classic double-bubble appearance on imaging. This is likely because since birth, our patient has been intolerant of enteral feeds, and was TPN dependent. She also has had a g-tube in place since 1 month of age. The lack of enteral intake and g-tube gastric decompression likely decreased the amount of duodenal dilation proximal to the atretic band, preventing the classical double-bubble presentation. It is acceptable to reason that this patient's diagnosis of duodenal atresia may have been made at an earlier timepoint if it had presented with the typical radiological findings commonly associated with the condition.

Our review of the literature revealed several cases of delayed diagnosis of duodenal anomalies, but no instance of duodenal atresia in a child as old as our patient. The oldest of these cases was a 16-year-old male with Down Syndrome, with 70% duodenal stenosis discovered after foreign body ingestion resulting in obstruction.[2] Another patient was a 5-month-old male with Down syndrome, who presented with hematemesis after previously tolerating breastfeeding without incident. Duodenal stenosis was diagnosed via a combination of upper endoscopy and upper gastrointestinal series, revealing some passage of contrast through a stenotic duodenum. Malrotation was also discovered intraoperatively in this patient.[3] A more severe case was of duodenal stenosis was discovered in malnourished 18-month-old boy, who presented with bilious emesis, failure to thrive, and a scaphoid abdomen.[4] Lastly, a 9-year-old with persistent vomiting was found to have a duodenal diaphragm on endoscopy.[5]

The vague clinical picture of disordered stooling, abdominal pain, bilious emesis, and intolerance of enteral feeding in a child has a broad differential diagnosis. In this case, workup was further complicated by the patient's concurrent medical history of MMHS. The transfer of patient care between multiple facilities provided an additional hurdle to overcome. This delay in diagnosis was the result of multiple confounding factors in a challenging case. The true source of the patient's symptoms was only elucidated when the diagnostic process was essentially reset, and anatomy was investigated under direct visualizing intraoperatively.

Conclusion

Duodenal anomalies should be considered in the differential diagnosis of patients presenting with persistent bilious

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emesis and obstructive symptoms, especially in the setting of inability to tolerate enteral feeding. Delay in diagnosis of duodenal anomalies may severely affect quality of life, as well as commit patients to invasive treatments, such as TPN dependence or surgical intervention. Missed diagnosis of a congenital anomaly should be contemplated in the systematic workup of atypically presenting or refractory patients.

Systematic review has identified that even common diseases are among missed diagnoses in admitted adult patients. At least 0.7% of hospitalized adults have been impacted by errors in diagnosis. As such, process improvement in the diagnostic course can be a point of potential intervention to mitigate these errors.[6] This concept can be extended to other patient populations, including pediatrics, to bolster the efforts of augmenting patient safety by encouraging thorough and thoughtful diagnostic processes.

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