

Duodenal Obstruction in Down's Syndrome Due to Malrotation and Duodenal Web

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ABSTRACT

This case reports on a child with Down's syndrome who had initial presentation of intestinal obstruction due to malrotation for which she went through correction via laprotomy and later found to have duodenal web with persistent vomiting after correction of the malrotation.

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Key words: Malrotation; Duodenal web; Down's syndrome; Persistent vomiting

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CASE REPORT

This is a 10 month old Asian female with Down's syndrome who presented to the emergency room with a history of projectile vomiting of coffee ground in appearance and was defecating black tarry stools.

She was born to a 44 years old Gravida 7 Para 7 at 38 weeks via normal spontaneous vaginal delivery with no post partum complications. She had tolerated regular cow's milk formula since birth. Three months prior to admission she was presented to the pediatrician's office with non projectile vomiting with couple of episodes of coffee ground emesis and 1 week history of bilious vomiting. Her stools were guaiac positive. Symptoms worsened when baby foods were introduced. She does have a history of intermittent constipation and required glycerin suppository

as needed. An upper GI series was ordered and revealed a high grade obstruction in the proximal duodenum with malrotation. The patient went for an exploratory laprotomy with correction of malrotation and appendectomy. She was discharged home in stable condition on a lactose free cow's milk protein formula.

One week after surgery when she developed intermittent episodes of projectile vomiting. She was seen in the emergency room and abdominal X-rays showed dilated bowel loops. She was referred to Pediatric Gastroenterology. On physical examination she was tachycardic but the rest of the vital signs were within normal limit. Weight at 10% and height at 25%. Abdominal examination revealed scar mark from previous exploratory laprotomy and abdomen was non distended and non tender. Bowel sounds were diminished. There was no evidence of organomegaly.

A repeat upper GI series was ordered. It revealed a partial obstruction in the second portion of duodenum. An esophageal gastroduodenoscopy was performed and revealed duodenal obstruction secondary to duodenal web at second portion of the duodenum. Laprotomy revealed a marked dilated first portion of the duodenum with a relatively collapsed small bowel distally. A duodenotomy was performed and the web was excised circumferentially. The web revealed a very small pin point opening. The ampulla of Vater was noted to the area distal to the web. Patient tolerated the procedure well. The nasogastric catheter was taken out on the seventh post operative day. Abdominal X-rays at early and late postoperative days were normal.

DISCUSSION

Congenital duodenal obstruction is one of the most common anomalies in newborns and infants. In 38-55% of patients, intrinsic duodenal obstruction is associated with another significant congenital anomaly^[1]. Duodenal web begin in the early embryologic development of the foregut. As the foregut undergoes lengthening and rotation, the hepatobiliary and pancreatic anlagen start as buds at the middle portion of the duodenum.

Other anomalies associated, such as intestinal malrotation and preduodenal portal vein, are well recognized. Superior mesenteric artery syndrome is a very rare condition in childhood most frequently resulting in duodenal obstruction in the adult population^[2]. Bleeding from ulcers at the diverticulum^[3], obstruction from gallstones^[4] and marbles^[5] have been described in different studies. Hepatic

hemangioma is an unusual finding in the pediatric population and is more common in older children and adolescents than in neonates and infants^[6,7]. About 10% of duodenal webs found in childhood are associated with other congenital conditions such as trisomy 21, intestinal malrotation, situs inversus, congenital heart disease, or annular pancreas^[8]. So far nearly 100 cases of duodenal web have been reported in the literature^[5,6,9]. The second part of the duodenum is the most common site, representing 85% to 90% of all duodenal web cases. The third and fourth parts of the duodenum represent 20% and 10% of duodenal web cases, respectively^[10,11].

This rare deformity is typically detected in childhood presenting with a gastric outlet obstruction. Delayed presentation of this lesion was reported in the older child and the adult population with nausea, vomiting, abdominal pain, weight loss, recurrent pancreatitis, and gastrointestinal bleeding^[11,12].

Clinical presentation may be characterized by nonspecific abdominal symptoms. Abdominal discomfort is usually located in the epigastric, right upper quadrant of abdomen or umbilical area, and is made worse or brought on by eating and relieved by vomiting, belching or assuming a certain posture^[6]. Late presentation could be due to perforated web, which allow food stasis and compensated mechanical gastric digestion without growth disturbance.

Diagnosis of duodenal web is quite difficult, even with some useful radiographical findings. Windsock sign or web or intraluminal duodenal diverticulum can be demonstrated during an upper GI series because a constant peristalsis in the proximal duodenum will cause the membrane to prolapse distally into the distal duodenum^[13].

Endoscopic resection of the duodenal membrane is not recommended due to the potential risk of the injury to the pancreatic or bile ducts^[13]. The reported experience of endoscopic treatment of duodenal webs in children is limited, possibly related to the inexperience most pediatric gastroenterologist typically includes dilation; however, the needs for laser therapy and cutting with a papillotome or sphincterotome are uncommon in children. The surgical treatment is therefore recommended. Both Heineke-Mickulitz-type (laprotomy) and laparoscopic duodenoduodenostomy have been performed to correct a duodenal web. The laparoscopic technique, a minimally invasive surgical technique, provides better visualization and easier approach whereas through standard laparotomy at right upper quadrant incision, it is more difficult to construct the anastomosis between the duodenal bulb and the small segment of distal duodenum^[14]. The disadvantage of the laparoscopic technique may limit the evaluation in the distal bowel segments because careful visual inspection of the bowel is more likely

to detect other obstructive lesions.

We aimed to present a case of Duodenal web with malrotation in the proximal duodenum in a patient with trisomy 21 which should be on a list of differential diagnosis in a patient with vomiting.

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