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Original article

Delayed presentation of congenital intrinsic duodenal obstruction in children with non-bilious vomiting: a diagnostic dilemma

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Abstract

The duodenum is described as the most common site of intestinal obstruction, classically presenting with bilious vomiting. Of the various categories described, congenital duodenal webs are reported as a rare cause of duodenal obstruction. The clinical features may vary depending on the size and location of the duodenal web. We are reporting 5 pediatric patients with delayed presentation of congenital intrinsic (type 1) duodenal obstruction. All patients presented with recurrent non-bilious vomiting and were misdiagnosed as gastroenteritis in other centers. The diagnosis was confirmed with upper gastrointestinal tract contrast studies. The patients were managed successfully with surgical intervention.

Keywords

Atresia, duodenum, non-bilious, stenosis.

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Introduction

Congenital duodenal atresia is rare, with a reported incidence of 1:10,000-40,000 births [1]. The intrinsic duodenal obstruction results from embryologic

defect during the development of the foregut, leading to defective recanalization [2]. The clinical presentation depends on the extent of obstruction (whether partial or complete) and usually manifests itself as intermittent or recurrent bilious or non-bilious vomiting and epigastric fullness. Complete duodenal atresia is usually symptomatic in the early neonatal period, while partial obstruction (i.e. duodenal web) may have late presentation and a more challenging diagnosis [3]. Although plain abdominal X-ray may be the most commonly used initial investigation, barium studies and flexible endoscopy of gastrointestinal tract are very helpful diagnostic modalities. Surgical treatment (either open or endoscopic approaches) is the first-line choice. The aim of this series is to highlight that even non-bilious vomiting should always be considered before ruling out this diagnosis.

Case details

Demographic and clinical details

We describe 5 patients (4 females) with mean age of diagnosis for duodenal obstruction of 2.8 years. At admission: anthropometric details were recorded and the following tests were performed: complete blood count (CBC), serum electrolytes, blood glucose levels, blood urea with serum creatinine, liver-specific aminotransferases (SGOT/PT) and urinalysis (**Tab. 1**).

Management

All the patients had been misdiagnosed with and treated for gastroenteritis in other centers; there was no fever or diarrhea/constipation. Multiple hospital admissions were reported for the same presentation. Patients were prescribed oral anti-emetics (domperidone/ondansetron) and needed intravenous fluids. However, the symptoms recurred 2-4 months after discharge. As outpatients, liver/kidney functions tests were normal, and random blood sugar level along with blood gas analysis was also found normal. One patient was submitted twice to antibiotic therapy for acute pulmonary disease (**Tab. 1**). One patient was investigated for celiac disease (TTG serology), and in another, neuroimaging (brain CT-scan) was performed to rule out cause of repeated emesis (nothing found). After a transient improvement with oral medications, the symptoms recurred, leading to referral of all the patients.

At our center, after adequate resuscitation, all patients started oral feeding, with recurrence of non-bilious vomiting. Chest X-rays and abdominal ultrasonography revealed unremarkable findings. Upper gastrointestinal tract contrast study was planned, which demonstrated grossly dilated stomach and duodenum (**Fig. 1**) with delayed passage of contrast distally. With a presumptive diagnosis of partial duodenal obstruction, patients were taken up for laparotomy. Per-operatively in each patient, a web with opening of variable size (type 1 atresia) was seen in the lumen of the second part of the duodenum, situated just proximal to ampulla. The web was excised and duodenoplasty was performed. In one patient, malrotation of the gut was found and was corrected by Ladd's procedure along with the duodenoplasty. The postoperative period was uneventful; oral feeding was started at first bowel movements; thereafter, the patients were discharged.

Discussion

Reported firstly by Boyd in 1845, congenital duodenal web (CDW) is a rare cause of duodenal

Table 1. Clinical and demographic details of the 5 patients.

No.	Age	Sex	Presenting complaints	Weight-for-height	Associated morbidities
1	3.5 Y	F	Recurrent non-bilious vomiting, failure to thrive, severe dehydration	< 3rd percentile	Anemia, electrolyte imbalance, pre-renal azotemia
2	2 Y	M	Recurrent non-bilious vomiting, early satiety, failure to thrive, recurrent chest infections	< 3rd percentile	Anemia, aspiration pneumonitis
3	2.8 Y	F	Recurrent non-bilious vomiting, loss of appetite, failure to thrive, pain in abdomen	< 3rd percentile	Anemia, per-operatively malrotation
4	3 Y	F	Recurrent non-bilious vomiting, lethargy, failure to thrive	< 3rd percentile	Hypoglycemia, deranged KFTs
5	2.5 Y	F	Recurrent non-bilious vomiting, poor tolerance to solid food, failure to thrive, cough	< 3rd percentile	Anemia, aspiration pneumonitis

Y: years; F: female; M: male; KFTs: kidney function tests.

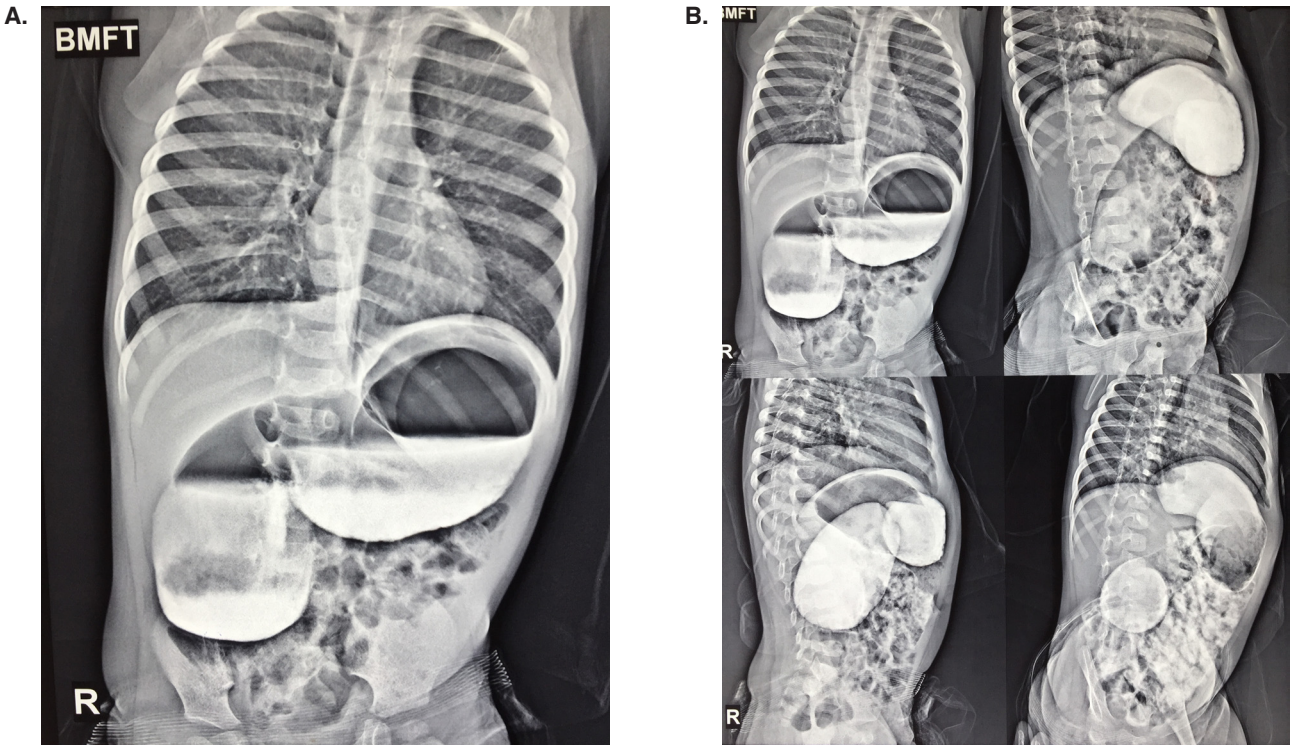

Figure 1. Gastrointestinal contrast study showing dilated stomach and duodenum.

obstruction [4]. Bilious vomiting is the most common presenting symptom, but non-bilious vomiting can occur in pre-ampullary obstruction. The timing and nature of clinical symptoms depend on the size of the opening in the web. It has been reported that the diagnosis of congenital duodenal obstruction (CDO) may be delayed even up to adolescence [5-7]. Mean age of diagnosis (33.6 months) in our series was much higher than the 9.5 months reported by Ratani et al. [8]. Mousavi et al. reported the mean age of presentation as 26.7 months in patients with partial duodenal obstruction [3].

The most important cause of delayed diagnosis in our series was the non-bilious nature of vomiting. Consequently, all our 5 cases had been treated for gastroenteritis in other centers. In partial obstruction, clinical presentation can be more confusing due to the association with nonspecific abdominal symptoms (such as epigastric discomfort, worsened by food intake and relieved after episodic vomiting) [9]. The association of CDW with other congenital disorders (i.e. Down syndrome, cardiac anomalies, malrotation of the gut, vertebral defects, renal anomalies, etc.) has been described, further complicating clinical presentation. Some studies have reported hematemesis as a presenting symptom of duodenal stenosis and atresia [10-12].

The diagnosis of CDO can be made prenatally by ultrasound screening [13]. Postnatally, an abdominal radiograph will typically show the characteristic 'double-bubble' sign in complete duodenal obstruction. In the case of partial obstruction, gastrointestinal contrast studies are considered the gold standard investigation. Flexible endoscopy can reveal overdistension of the duodenum, the protrusion of the duodenal web and, if any, the mucosal diaphragm in the stenotic lumen of the duodenum [14]. The presence of recurrent vomiting, along with systemic signs (like failure to thrive, anemia and malnutrition), should be taken as a marker of some serious disease and warrants adequate investigational work-up.

Surgical procedures (duodenoplasty or duodeno-duodenostomy and dudeno-jejunostomy) are commonly used for the treatment of CDW [15, 16]. Recently, endoscopic surgery with a high-frequency wave cutter or a balloon catheter has been reported to be a safe and effective intervention [17]. Laparoscopically assisted management has also been used as therapeutic intervention, but all these newer approaches require great expertise and are not always available in developing countries [18].

Conclusion

Although rare in terms of incidence, CDW with aperture can present variously as to the extent of the intrinsic obstruction. High index of suspicion,

detailed anamnesis, good clinical examination and appropriate investigation work-up in a patient with recurrent non-bilious vomiting can assist timely diagnosis.

Declaration of interest

The Authors declare that they have no competing interests. This research did not receive any specific grants from public, commercial, or non-profit funding agencies.

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