GUESS THE DIAGNOSIS

Upender Shava[1], Rishi Bolia[2], Zaheer Nabi[1], Reddy D N [1]
1- Division of Pediatric Gastroenterology, Asian Institute of Gastroenterology, Hyderabad
2- Division of Pediatric gastroenterology, Postgraduate Institute of Medical Education and Research, Chandigarh

Case

A 5-year-old girl presented with complaints of vomiting since infancy. She was having bilious vomiting one to two times per week with increased frequency of vomiting over the last 1 years. She also complained of pain and upper abdominal distension, which used to decrease after vomiting. On examination she had significant failure to thrive and her abdominal examination revealed epigastric distension. Investigations revealed normal complete blood count, serum creatinine, and thyroid profile. Abdominal ultrasonography was normal. Her abdominal x-ray (erect) and barium meal follow-through showed the following (Figure 1 & 2) –

What is the most likely diagnosis?

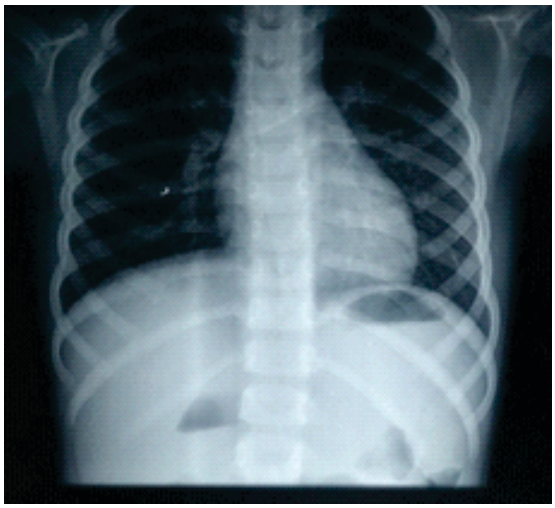

Figure 1

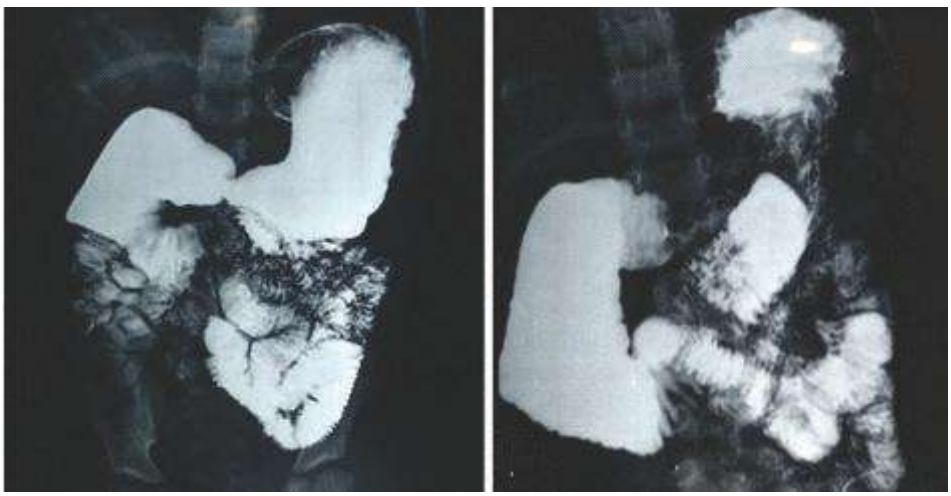

Figure 2

Answer

Incomplete congenital duodenal obstruction, most likely a Congenital duodenal web (CDW).

Follow – up and discussion

Her abdominal x-ray (erect) showed a double bubble sign and barium meal follow-through revealed narrowing in the third part of the duodenum with a dilated proximal duodenum. The appearance was a classical wind sock web deformity. Subsequently, an UGI endoscopy was performed which showed dilation of the first and second part of duodenum and narrowing with a tiny lumen in the third part of duodenum. (Figure 3) confirming the diagnosis of a congenital duodenal web. The patient underwent surgery (duodeno-duodenostomy) for the same.

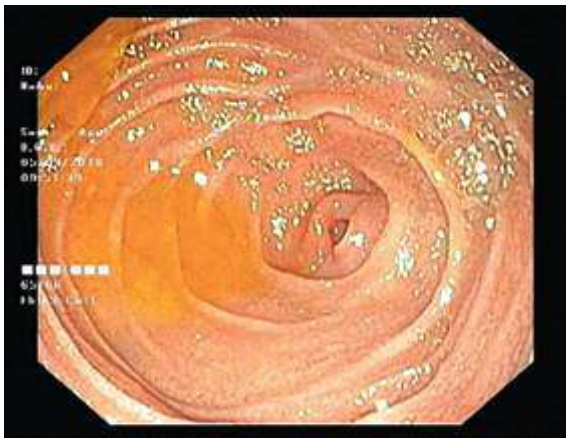

Figure 3

Congenital duodenal web (CDW) is a rare condition (1:10,000 to 1: 40,000) that usually presents early in life. It was first reported by Boyd in 1845. It is believed to develop secondarily to a failing in recanalization of the duodenal lumen between the 8th and 10th weeks of gestation. The failure of recanalization leaves behind a thin web with mucosa and submucosa layers only; the muscular layer is absent.

Patients with complete-type duodenal web present early in the neonatal period with duodenal obstruction whereas the fenestrated type may present late. Reports also exist of congenital duodenal web diagnosed in adulthood The second part of the duodenum (distal to the ampulla), is the most common site (1) This is in contrast to an annular pancreas which is most frequently pre – ampullary. Disorders like Down syndrome, cardiac anomalies, malrotation of the gut, vertebral defect, renal anomalies etc. have been described as an association in upto 50% of patients with CDW (2) Our patient did not have any of the above.

Surgery, in the form of excision and duodenoplasty or bypass procedures like duodeno-duodenostomy used to be the conventional modality for treatment of CDW (3) However, in recent years' endoscopic procedures such as endoscopic membranotomy with sphincterotome or needle knife and endoscopic balloon dilatation have been reported (4,5)

In conclusion, CDW should be suspected in cases of abdominal distension with bilious vomiting in children, even those who are relatively older. Endoscopy usually confirms the diagnosis. Treatment has traditionally been surgical however endoscopic modalities have emerged as a therapeutic option in recent years.

References

1. Beeks A, Gosche J, Giles H. et al. Endoscopic dilatation and partial resection of a duodenal web in an infant. J Pediatr Gastroenterol Nutr. 2009;48:378–381.
2. Sarin Y K, Sharma A, Sinha S. et al. Duodenal webs: an experience with 18 patients. J Neonatal Surg. 2012;1:20.
3. Mustafawi A R, Hassan M E. Congenital duodenal obstruction in children: a decade's experience. Eur J Pediatr Surg. 2008;18:93–97
4. DiMaio C J, Kamal N, Hogan C M. et al.Pediatric therapeutic endoscopy: endoscopic management of a congenital duodenal web. Gastrointest Endosc. 2014;80:166–167
5. Poddar U, Jain V, Yachha SK, Srivastava A. Congenital duodenal web: successful management with endoscopic dilatation. Endosc Int Open. 2016 Mar;4(3):E238-41.