Jejunal web masquerading as intestinal malrotation in a neonate with bilious vomiting: A case report


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Abstract
Neonatal bilious vomiting is considered as a pathological sign and warrant urgent medical attention. Upper gastro-intestinal contrast study can be notoriously difficult to diagnose incomplete intestinal rotation, especially when there is partial hold up of the contrast at the duodenum. We report a case of a full term neonate with incomplete jejunal obstruction due to a fenestrated jejunal web, presenting with bilious vomiting and masquerading as intestinal malrotation in the contrast study. We discuss the recent literature and the potential diagnostic difficulty in the diagnosis of jejunal web. In view of the low specificity of radiological imaging study of jejunal web, clinicians should be aware that congenital gastro-intestinal webs are a possible cause of partial gastro-intestinal obstruction in infants and children.

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Bilious vomiting is often considered a pathological sign in a neonate and would prompt urgent medical attention. Differential diagnosis for bilious vomiting includes severe sepsis, functional gastro-intestinal obstruction such as long segment Hirschsprung's disease, anatomical gastro-intestinal obstruction such as intestinal malrotation and intestinal atresia. Type 1 intestinal atresia, according to the Louw and Barnard classification [1] is defined as an intra-luminal web resulting in complete or incomplete intestinal obstruction, it is most commonly found at the duodenum and would classically give rise to the double bubble sign on plain abdominal X-Ray. However, when the intestinal obstruction is incomplete owing to a fenestrated membrane, the diagnosis can often be delayed and confused with other causes of partial intestinal obstruction. We report a case of a neonate presenting with bilious vomiting due to an incompletely obstructing jejunal web masquerading as malrotation and we review the recent literature on this uncommon clinical entity.

1. Methods

Baby boy L was born full term with a birth weight of 3.15 kg. He has unremarkable antenatal history and there was no maternal history of polyhydraminos. He presented to our hospital with repeated vomiting with undigested milk on day 7 of life. He had normal meconium passage on first day of life and has normal daily bowel opening. Clinical examination revealed a mildly dehydrated neonate, soft, non-distended abdomen with a 6% drop in body weight and blood test was unremarkable with normal electrolytes. Plain abdominal X-ray (Fig. 1a) showed distended stomach and duodenum, distal bowel gas pattern was unremarkable. After admission to the ward, he started to develop bile stained vomiting. An oral gastric tube was inserted yielding large amount (20 ml) of bilious aspirate. His temperature remained stable and sepsis markers were not raised. In view of the clinical suspicion of malrotation with volvulus, urgent upper gastro-intestinal contrast study was arranged on the same day and it showed incomplete duodenal obstruction, distended duodenum with partial hold up of contrast at the distal duodenum. Duodeno-jejunal junction was at the midline of the spine (Fig. 1b). Incomplete intestinal rotation with partial duodenal obstruction and impeding volvulus was considered as the top differential diagnosis.

2. Results

Emergency laparotomy performed and showed normal left sided duodeno-jejunal junction, no Ladd's band and no intestinal malrotation. However, there was grossly distended duodenum and proximal jejunum correlating with the contrast findings, with a
transition zone at 7 cm distal to the duodeno-jejunal junction. Upon longitudinal enterotomy at the distended proximal jejunum, a 5 French infant tube was passed distally to confirm the presence of a jejunal web when it encountered resistance at the site of the web, correlating with the palpable fibrotic ring extra-luminally (Fig. 2). The jejunal web together with 2 cm of the jejunum was excised and a primary end to oblique jejuno-jejunostomy anastomosis was performed with 5/0 PDS interrupted. Luminal patency of the distal bowel was confirmed with normal saline injection all the way to rectum. The post op recovery was uneventful. And full oral diet was resumed and tolerated. The baby was discharged on the thirteenth day post op. Pathological report of the resected jejunum showed granulation tissue proliferation with serosal fibrosis. Further medical screening of the baby did not reveal any cardiac, renal or skeletal anomalies.

3. Discussion

We reported an uncommon clinical finding of a type 1 intestinal atresia at the proximal jejunum presenting as bilious vomiting in a full term newborn baby. His initial clinical presentation of bilious vomiting and radiological findings closely resemble that of an intestinal malrotation.

Neonatal bilious vomiting without abdominal distension remains a surgical emergency in newborn and requires urgent attention. Intestinal malrotation must be urgently excluded with an upper gastro-intestinal contrast study. However, a false positive rate of 15% was reported with the diagnosis of malrotation on the basis of an upper GI series, even to an experienced radiologist [2]. This is especially true when it can present as a spectrum of varying degree of incomplete intestinal rotation to complete malrotation. A normal plain abdominal X-ray in infants does not rule out malrotation. Sometimes, the contrast study may reveal dilated loops of bowel, suggestive of intestinal web or mimicking a bird’s beak appearance consistent with volvulus. A partial intestinal obstruction due to a fenestrated intestinal web are often difficult to diagnose.

Congenital web of the gastro-intestinal tract is an uncommon anomaly causing GI obstruction in infants. The most common site of intestinal web is the second portion of duodenum [3], and it would often be associated with trisomy 21, cardiac or renal anomalies, and clinically it would usually present as double bubble sign on the abdominal X Ray and non-bilious vomiting, depending of the site of obstruction in relation to the ampulla of Vater. Windsock deformity of the jejunum due to congenital web is a rare anomaly with scant literature. In the case reported by Andrews and Stem [4], the baby with jejunal web presented with small bowel obstruction and bile-stained vomitus, and she was treated with excision of the web and jejunoplasty. Multiple jejunal web could also be found in some instances [5], resulting in type IV jejunal atresia, therefore it is essential to ascertain the luminal patency of the distal bowel after enterotomy.

In the review by Lin et al. from Taiwan, from the 37 congenital webs of the gastro-intestinal tract in children in the past 20 years, only 8% of the webs arise from the jejunum [6]. And the presenting symptoms of such webs can often be vague and non-specific, especially when the fenestrated mucosal web results in partial obstruction. Symptoms include vomiting, abdominal distension,
failure to thrive, volume depletion and poor weight gain, which may represent a sub-acute or chronic condition. Diagnosis of an incompletely obstructing web can be difficult and the mean time to diagnosis for those with jejunal webs was quoted as 231 days [6]. In our case, the initial confusion of intestinal malrotation by the imaging study prompted urgent laparotomy, leading to a much earlier operative diagnosis before the development of further complications.

The etiology of intestinal atresia remains unknown. According to the recanalization theory proposed by Tandler [7], the duodenal endoderm thickens and obliterates the lumen before it recanalizes. Another widely adopted theory by Louw and Barnard [1] suggested intrauterine vascular events causing disruption of mesenteric vessels and subsequently intestinal atresia. Recently, there has been case report suggesting intestinal hyper-proliferation and mucosal hyperplasia to be a cause for jejunal web in an infant [8].

Given the low specificity of radiological imaging study for jejunal web, we suggest that clinicians should have a high index of suspicion and be aware that congenital GI webs are a possible case of GI obstruction in infants and children.

References