Case Report

Simultaneous duodenal stenosis and duodenal web in a newborn

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Abstract

Failure of duodenal recanalization results in a spectrum of proximal bowel obstruction from stenosis to atresia. Associations between congenital duodenal obstruction and other congenital anomalies have been well documented although the coincidence of duodenal stenosis and duodenal web is incredibly rare, posing a unique diagnostic challenge. We report a case of a full-term 4-day-old female child presented with forceful, bilious emesis and poor oral intake with decreased frequency of urination, and stooling whose initial abdominal radiograph showed several loops of gas-filled bowel in the distal stomach and proximal duodenum mimicking the classic “double-bubble” sign. An upper gastrointestinal barium contrast study revealed distention of the duodenal bulb with an abrupt narrowing and subsequent dilation at the second portion of the duodenum raising the suggestion of multiple duodenal obstructions. Ladd’s procedure was performed, and the stenotic and webbed segments were bypassed with a Kimura diamond-shaped duodenoduodenostomy. © 2016 the Authors. Published by Elsevier Inc. under copyright license from the University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Case presentation

A full-term 4-day-old female child presented with forceful, bilious emesis and poor oral intake with decreased frequency of urination and stooling. The mother had received routine prenatal care, and screening laboratories were unremarkable with the exception of positive group B streptococcus culture, which was adequately treated with intrapartum penicillin prophylaxis. Prenatal ultrasound revealed a 2-mm cystic structure believed to represent a benign cyst or bowel gas.

Physical examination of the infant on presentation demonstrated a firm abdomen without significant distention or palpable masses. The anus was patent, and the infant had been stooling well before admission. An abdominal radiograph showed several loops of gas-filled bowel in the distal stomach and proximal duodenum mimicking the classic “double-bubble” sign (Fig. 1). An upper gastrointestinal contrast study performed at the time of admission revealed distention of the duodenal bulb with an abrupt narrowing and subsequent dilation at the second portion of the duodenum. A small amount of contrast was present in the nondistended...
Duodenal stenosis is the result of incomplete duodenal lumen recanalization, occurring sometime around gestational week 7 [2]. The exact pathogenesis of this condition remains unclear, although rare cases in siblings have been reported suggesting a possible genetic predisposition [3]. The prevalence of duodenal atresia is approximately 0.9 per 10,000 live births according to recent estimates, which has remained essentially stable over time [4]; less data exist on the incidence of duodenal stenosis. Duodenal webs, a similar condition, are the result of incomplete bowel lumen recanalization occurring between the eighth and tenth weeks of gestation [2]. Less epidemiologic data are available regarding duodenal webs; however, they are thought to be more rare with an estimated incidence somewhere between 1 in 10,000 and 1 in 40,000 [5]. Associations between congenital duodenal obstruction and other congenital anomalies have been well documented. Down’s syndrome is the most common, but association with other conditions have been reported including gut malrotation, imperforate anus, cardiac malformation, and annular pancreas [2,6].

Double-duodenal obstructions similar to the one described in this case have been reported in the literature, although these cases have been mostly anecdotal. To the best of our knowledge, there have been 31 cases of intrinsic double-duodenal obstruction reported since Boyd first described the finding in 1845 [3,7-10]. Reid’s [11] study of 140 patients with intrinsic duodenal obstructions noted only 4 instances with multiple obstructions.

The classic radiographic finding associated with duodenal obstruction is the “double-bubble” sign on plain film x-ray. This finding is best visualized in cases of duodenal atresia or severe stenosis in which the proximal, left-sided bubble represents air and fluid in the stomach and the distal, right-sided bubble represents air in the dilated segment of the proximal duodenum [12]. Variations of this radiographic finding may be seen including the presence of distal bowel gas depending on the degree and location of obstruction [13]. Ultrasound can also be used in the antenatal period to raise suspicion of duodenal obstruction by demonstrating fluid filled structures in the upper abdomen with a paucity of fluid distally [12]. Suspicious sonographic findings of this nature during the prenatal period should be interpreted with caution as they may simply represent duodenal fluid accumulation secondary to slow peristalsis [12].

Radiographic findings in patients with double-duodenal obstruction have been quite varied. Some cases have presented with the classic “double-bubble” sign on plain abdominal films [9,14]. Stringer et al [15] reported on 4 patients with double-duodenal obstruction, two of which had atypical plain abdominal films including calcification and abnormal gas patterns suggestive of more complex pathology. Prenatal ultrasound performed on one patient later found to have double-duodenal atresia demonstrated 3 isolated cystic structures in the upper abdomen [16].

Fig. 1 – An abdominal radiograph demonstrates 3 air-filled portions of bowel in the region of the distal stomach and proximal duodenum. Distal air indicates an incomplete obstruction.

Congenital duodenal obstructions represent a spectrum of fairly uncommon disorders with the potential for devastating consequences if not properly managed in an expedient fashion. While many cases present with the classic “double-bubble” sign on plain film x-ray, findings on radiographs, and other imaging modalities may vary. In the described case, a rare double-intrinsic duodenal obstruction was discovered despite seemingly benign findings on prenatal ultrasound. Therefore, it may prove extremely beneficial to closely follow newborns with similar sonographic findings in the immediate postnatal period. Furthermore, maintaining a high index of clinical suspicion is critical for properly diagnosing any duodenal obstruction in the acute setting.
Fig. 2 – Fluoroscopic upper gastrointestinal study. Contrast outlines 2 distended lumens, the first being the proximal duodenal bulb (arrows), and the second being the second portion of the duodenum (dashed arrows) on frontal and lateral views. Note the normal caliber fourth portion of the duodenum (arrowheads).