Case Report

Y- Shaped Sigmoid Duplication in a Newborn: Report of a Rare Congenital Anomaly

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Abstract
Y-shaped sigmoid duplication is very rare congenital anomaly. Only few cases have been reported in English literature. Our patient, 19 days female, presented with abdominal distension, constipation and bilious vomiting. She was asymptomatic for initial 9 days of life. The abdomen was grossly distended and a large lump was palpable. Plain X-ray abdomen showed a large air collection. On exploration, Y-shaped duplication was found arising from mid sigmoid. The other end was communicating with a large cyst filled with fecal matter. Duplicated segment was excised with simple closure of sigmoid.

INTRODUCTION
Intestinal duplications are rare congenital anomalies with an incidence of 1 in 4000–5000 births [1]. Small bowel is the most common site and colonic duplications represent less than 10–15% of all intestinal duplications [1,2]. Y- Shaped duplication is extremely rare form of tubular colonic duplications. We report a neonate with Y-shaped sigmoid duplication who underwent successful treatment.

CASE PRESENTATION
A newborn girl, product of full-term normal delivery, was admitted at 19 days of life with gradually increasing abdominal distension and worsening constipation for duration of 10 days. There were no positive findings reported on antenatal scans. She also had several episodes of bilious vomiting since 5 days. The child passed meconium within 24 hours of birth and was passing stools normally till day 9 of life. Following this the frequency and amount of stools decreased gradually and there was no passage of stools since 5 days. The child was given formula feeds in addition to breast milk from day 1 of life. There was no history of fever and loose stools.

On examination, the baby was dehydrated but hemodynamically stable. The abdomen was grossly distended. There were no signs of peritonitis. A large, intra-abdominal lump was palpable occupying almost whole of the abdomen only sparing left hypochondrium and left iliac fossa. Bowel sounds were increased and were heard only on left side of abdomen. Perineal examination revealed normal external genitalia and anal opening.

On plain x-ray abdomen, a large air collection was present on right side, extending up to midline, stomach was distended and bowel loops were seen on left side (Figure 1).

Differential diagnosis of Neonatal Enterocolitis (NNEC) with contained perforation, hirschprungs disease and segmental bowel dilatation were kept before exploration.

In view of definite intestinal obstruction and gross abdominal distension, no further investigations were performed. After initial resuscitation with IV fluids, the child was taken up for exploratory laparotomy. Laparotomy was performed from right upper quadrant incision. A large cystic lesion was present occupying more than two-third of abdomen. Ascending colon was compressed anteriorly on the cyst (Figure 2a) and rest of the bowel was healthy and was pushed towards left. The cyst was opened (Figure 2b) as there was no space for manipulation in abdomen and it revealed large amount of fecal matter. Any communication of cyst with bowel could not be identified at right side, extending up to midline, stomach was distended and bowel loops were seen on left side (Figure 1).

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Figure 1 X-ray abdomen showing large air collection on right side of abdomen, distended stomach and bowel loops on left side.
that point of time. On further exploration, we found Y-shaped duplication arising from mesenteric side of sigmoid colon with narrow communication at mid-sigmoid (Figure 2c). The length of tubular segment was 5 cm (Figure 2d) and other side was ending at posterior wall of the cyst with a pinpoint communication. The cyst was partially excised and at remaining places, mucosa was stripped off. Abdomen was closed after lavage and putting a glove drain in the cyst cavity.

The child remained stable in postoperative period. There was purulent discharge from drain site which gradually decreased in amount and stopped in a week time. She was orally allowed on postoperative day (POD) 5 and was discharged on POD 9 on full oral feeds. The patient is doing well at 2 months postoperatively.

**DISCUSSION**

Colonic duplications are rare congenital anomalies. Tubular colonic duplications represent only 10% [3] and usually located parallel to the native colon. Y-shaped duplication is an exceptional form and only few cases of Y-shaped sigmoid duplications are reported in English literature [4-7]. There was female preponderance with age ranging from newborn to 35 years. Presenting symptoms were pain, abdominal mass [4,6] and features of obstruction [4] in older patients whereas newborns presented with perforation peritonitis [5] and abdominal distension [7]. Duplication was an intraoperative finding in patient with perforation [5] whereas other patients were diagnosed on lower gastrointestinal series [4,6,7]. Only one patient [7] had cystic component at the distal end of tubular segment like our patient.

These lesions have communication with bowel at one end and no communication at other end, so fecal matter and air accumulates because of inadequate drainage. The patients usually present with pain, lump or features of intestinal obstruction. Our patient was passing stools normally up to 9 days of life and developed features of obstruction when the terminal part of duplication accumulated a large amount of fecal matter and air and compressed ascending colon.

The duplication may be associated with vertebral, spinal, genitourinary and cardiovascular anomalies. There was no associated anomaly in our case.

The condition is rarely diagnosed accurately before surgery, as in our case we did not think of duplication. We kept the possibilities of NNEC with contained perforation, Hirschsprung disease and segmental bowel dilatation. NNEC with perforation and pseudocyst formation was the most probable diagnosis because the child was given formula feeds. The other differential of Hirschsprung disease with grossly dilated sigmoid was kept but colon will perforate before reaching to those dimensions and the lump is unusual in that situation. The other possibility of segmental bowel dilatation was also kept. Contrast enema would have been helpful in making the diagnosis preoperatively but it was not done due to grossly distended abdomen.

Double-barreled duplications require resection of bowel with end to end anastomosis whereas Y-shaped duplications can be resected completely without compromising the blood supply of native bowel as in our case.

In summary, although rare, a possibility of duplication should be kept in mind in patients presenting with lump, progressive constipation and large gas shadow on plain abdominal x-ray.

**REFERENCES**


