Case Report

Congenital Segmental Dilatation of Jejunum: A Rare Entity

Amar A. Shah, Anirudh V. Shah

Neonatal and Pediatric Surgeon, Amardeep Multispeciality Children Hospital and Research Center, Ahmedabad, Gujarat, India

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Abstract

Segmental dilatation of the intestine is a rare disease and mostly involves the ileal segment. It commonly presents in the neonatal period and early infancy with symptoms of partial or total obstruction. We report a rare case of an isolated jejunal segmental dilatation in a 7-year-old girl. The child presented with malnutrition and signs of subacute obstruction. The diagnosis was confirmed intraoperatively, and the dilated segment was resected. Although the etiology remains unknown, we discuss its clinical aspects and relevant literature.

Keywords: *Dilatation, jejunum, segmental*

Introduction

Segmental intestinal dilatation is a rare anomaly which can involve any part of the bowel. However, ileal involvement is most common. The histology of the dilated segment usually shows normal neurological innervation with a hypertrophied or thin muscle layer. There is no known abnormality of the enteric nervous system and pacemakers. The etiology remains unknown, and resection of the involved segment is the only treatment option.

Case Report

A 7-year-old girl presented with a complaint of recurrent abdominal pain for the past 4 years. She was born at term by vaginal delivery and had a normal neonatal period. She was immunized for her age and on symptomatic treatment till presentation. For the past year, she suffered from loss of appetite and had nonbilious vomiting 10–15 min after meals. The frequency of abdominal pain also increased over the preceding 6 months. On examination, the child was anemic (Hb 7.6 g/dl) and malnourished. Her abdomen was full but soft. She had a large palpable loop of bowel over the central abdomen. There were no signs of peritonism. Plain X-ray of the abdomen showed a large fluid level in the epigastrium with small fluid levels over the central abdomen. Ultrasound showed an overdistended stomach with the cecum and the ileocecal junction in the left hypochondrium. Upper gastrointestinal contrast study revealed a distended stomach and a dilated, redundant jejunal loop. Only a small amount of contrast was seen passing beyond this jejunal segment even after 4 h of the study [Figure 1].

The child underwent an exploratory laparotomy. At surgery, 12 cm of the jejunal segment which was 5 cm distal to the DJ flexure, was massively dilated [Figure 2]. The bowel proximal and distal to the dilated segment was normal. The dilated segment was resected and end-to-end jejunojejunal anastomosis was done after mobilizing the DJ flexure. The postoperative recovery was uneventful. At 6-month follow-up, she had gained 5 kg weight.

The histopathology of the resected segment showed thinning of the muscle coat with focal inflammation and no heterotopic tissue [Figure 2]. Ganglion cells were present all throughout the specimen.

Discussion

Around 150 cases of segmental intestinal dilatation are reported in the literature, but none of them provide any clues to the definite etiology of this disease.[1] It often manifests as an isolated, dilated small bowel segment without evidence of intrinsic or extrinsic obstruction or abnormal neural innervation. In the neonatal period, it may present with acute intestinal obstruction or can

Address for correspondence: Dr. Amar A. Shah, Amardeep Multispeciality Children Hospital and Research Center, 65, Pritamnagar Society, Near Government Ladies Hostel, Near Gujarat College, Ellisbridge, Ahmedabad - 380 006, Gujarat, India.
E-mail: shahamar22@gmail.com

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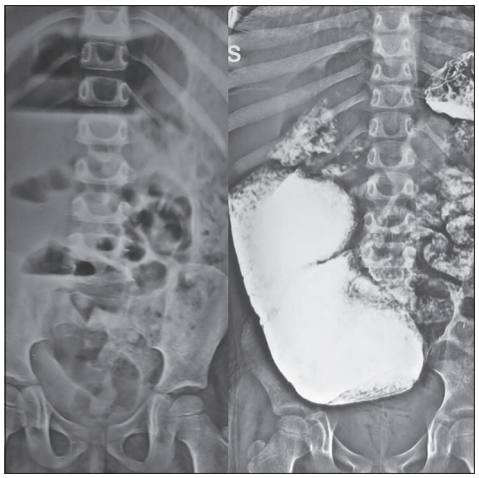

Figure 1: Plain X-ray abdomen showing a dilated stomach with scattered small air fluid levels on the right of the abdomen. Contrast study at 4 h showing the dilated and redundant jejunal loop with some contrast having gone into the ileal loops

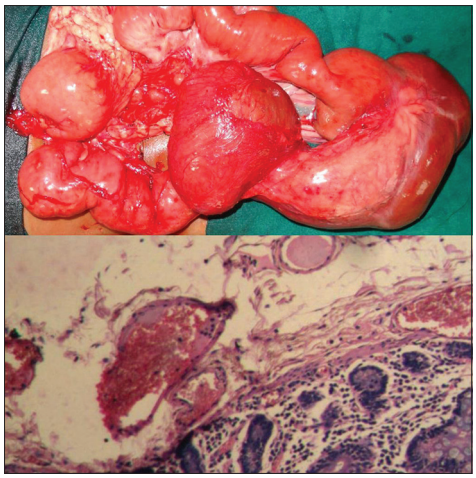

Figure 2: Peroperative photograph showing segmental dilatation of the jejunal loop distal to the DJ flexure (above) and thinning of muscle coat (H and E, ×10; below)

mimic volvulus or Hirschsprung's disease.[2] In older infants, it presents with anemia, malabsorption, chronic constipation, features of intussusception, or intermittent intestinal obstruction.[3] Uncommon presentations may be in the form of recurrent abdominal pain, bowel obstruction, and incidentally detected enterolith. Few may remain asymptomatic and present in their late adulthood. In a review of 36 cases by Ratcliffe *et al*., 50% of the cases had associated gastrointestinal tract anomalies, of which seven cases had Meckel's diverticulum, six had omphalocele, and six had malrotation of the gut.[4] Swenson and Rathauser in 1959 established the criteria for the diagnosis of this rare entity.[5] Their criteria included (i) limited bowel dilatation with a 3-fold to 4-fold increase in size, (ii) an abrupt transition between dilated and normal bowel, (iii) no intrinsic or extrinsic barrier distal to the dilatation, (iv) clinical picture of intestinal occlusion or subocclusion, (v) a normal neuronal plexus, and (vi) complete recovery after resection of the affected segment.

The presence of heterotopic tissues such as lung, pancreatic, esophageal, gastric, cartilage, and striated muscle in the dilated segment has been described by some authors.[6] Partial or complete absence of muscularis propria in the dilated segment has also been reported.[7] Some authors suggest intrauterine vascular accidents or external compression to the fetal bowel as a probable cause of the intestinal dilatation. Cheng *et al*. demonstrated localized vacuolization of the intestinal smooth muscle in their case, suggesting myopathy to the cause of dilatation.[8] Okada *et al*. suggested disorders of the interstitial cells of Cajal as a contributing factor for localized myopathy contributing to the segmental dilatation of the intestine.[9] Some authors suggest ganglionic dysplasia as a cause of congenital segmental dilatation.

Although segmental dilatation can involve anywhere from duodenum to distal colon, ileum is the most commonly affected site. In this case, the involvement of jejunum makes it more uncommon. The hypomotility in the involved segment caused the intestinal contents to gradually pool up. This led to progressive dilatation of the duodenum and the stomach, pooling of the intestinal contents therein, and malnutrition. It was only when the overdistended stomach resulted in persistent vomiting that medical attention was sought.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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