Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Jejunal duplication cyst in a female neonate: a case report

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Introduction and importance: Duplications are the abnormal portion of the intestine, either externally attached to the intestine or intrinsically placed within the bowel lumen. Their prevalence is noted to be around one in 25 000 deliveries. The rare gastrointestinal tract duplication may be located in any part of the gastrointestinal system from the oral cavity to the anus. The most common site of enteric duplication cyst (DC) is the terminal part of the ileum. Hence, duplications in jejunum are rare.

Case presentation: Hereby, the authors report a case of jejunal DC in a female neonate which was managed successfully via surgery and adequate post-operative care without any complications.

Clinical discussion: Duplications are more frequently single. They are usually located in the mesenteric border of the associated native bowel and may vary in shape and size. Most of them are cystic, followed by tubular and mixed type, with or without other congenital anomalies. More than 80% of the cases present before the age of 2 years as an acute abdomen or bowel obstruction, but many duplications remain silent unless complications occur, and therefore may not be diagnosed until adulthood. Complications of enteric DC include volvulus, bleeding, and, rarely, malignant degeneration.

Conclusion: It is important for paediatric surgeons to include DC in the differential diagnosis if a neonate presents with features of intestinal obstruction.

Keywords: cystic, duplication, gastrointestinal tract, proximal jejunum, surgery

Introduction

Duplications are the abnormal portion of the intestine, either externally attached to the intestine or intrinsically placed within the bowel lumen[1–4]. Their prevalence is noted to be around one in 25 000 deliveries[5]. The rare gastrointestinal tract duplication may be located in any part of the gastrointestinal system from the oral cavity to the anus[6]. The most common site of enteric duplication cyst (DC) is the terminal part of the ileum[7]. Duplications are more frequently single. They are usually located in the mesenteric border of the associated native bowel and may vary in shape and size. Most of them are cystic, followed by tubular and mixed type, with or without other congenital anomalies[8]. More than 80% of the cases present before the age of 2 years as an acute abdomen or bowel obstruction, but many duplications remain silent unless complications occur, and therefore may not be diagnosed until adulthood[8]. Complications of enteric DC include volvulus, bleeding, and, rarely, malignant degeneration[9]. Hereby, we report a case of jejunal DC in a female neonate which was managed successfully via surgery and adequate post-operative care. This case report has been reported in line with the SCARE criteria 2020[10].

HIGHLIGHTS

- Duplications are the abnormal portion of the intestine, either externally attached to the intestine or intrinsically placed within the bowel lumen.
- The rare gastrointestinal tract duplication may be located in any part of the gastrointestinal system from the oral cavity to the anus.
- The most common site of enteric duplication cyst is the terminal part of the ileum.
- Most of them are cystic, followed by tubular and mixed type, with or without other congenital anomalies.
- Jejunal duplication cyst is rare.
- This is the first case report of its kind to be reported from Nepal.

Case presentation

A single live female, small for gestational age, was delivered via emergency lower segment caesarean section at 38 weeks + 1 day

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Annals of Medicine & Surgery (2023) 85:5724–5727

Received 18 June 2023; Accepted 5 September 2023

Published online 14 September 2023

http://dx.doi.org/10.1097/MS9.0000000000001303

period of gestation. On the fifth day after birth, the infant presented to the paediatric emergency department with the chief complaints of increased sleep, decreased feeding, abdominal distension, and fever for 4 days. The mother reported that the infant was apparently well 4 days prior when she noticed increased sleep with no abnormal body movements and decreased feeding and abdominal distension. The infant had a history of fast breathing. There was no history of vomiting, diarrhoea, loss of consciousness, or abnormal body movements. The infant had a fever with a documented temperature of 102°F that was acute in onset, and no medication was given for it.

The infant was born via lower segment caesarean section due to failed induction at 38 weeks + 1 day period of gestation. The birth weight was 2300 g. The ultrasound before the delivery also revealed a single live intrauterine pregnancy of average gestational age of 37 weeks + 1 day in cephalic presentation with dilated bowel loops in intrauterine ultrasound measuring up to 13.9 mm seen in the bilateral lower abdominal cavity (as shown in Fig. 1). There was no significant family history. The mother was 33 years old, with a history of hypothyroidism, primary subfertility conceived via induction of ovulation, and polycystic ovary syndrome. The mother had regular antenatal visits, was immunized, and had no history of vaginal bleeding, leaking, fever, rashes, or lymphadenopathy. The mother had no history of gestational hypertension or gestational diabetes.

On examination, the infant was lethargic, had laboured breathing, and suprasternal retractions were present. The heart rate was 180 beats per minute, respiratory rate was 52 breaths per minute, oxygen saturation was 98%, the temperature was 98°F, and capillary refill time was less than 3 seconds. The central nervous system examination showed diminished Moro's reflex, Rooting reflex, and sustained suckling. The cardiovascular system examination was normal. Chest examination showed bilateral air entry with audible grunts. Abdominal examination revealed a soft, non-tender, non-distended abdomen. The anterior fontanelle measured 0.5×0.5 cm. Continuous monitoring was done for the need for intubation. Abnormal tightening of both upper limbs was noted with tachypnea, for which an injection Levetiracetam was given. Lumbar puncture was done under all aseptic conditions. The cerebrospinal fluid examination was normal, and meningitis was ruled out. Injection of Ciprofloxacin and Gentamicin were started.

Ultrasound was planned, which showed some prominent dilated bowel loops, the largest measuring 2.36 cm. Surgical consultation was done for the nasogastric drain, and antibiotics were escalated to (Piperacillin + Tazobactam), Vancomycin, and Metronidazole in view of gastrointestinal infection. Chest X-ray showed findings suggestive of pneumonia. The infant progressed to respiratory distress (Downes 3/10) with excessive secretion from the nasogastric tube, amounting to about 18 ml/day. Urine output was normal. On the following day, the NG drain was increased to 77 ml. ON *P*/A examination, soft liver-4 cm below the right costal margin and spleen was palpable suggesting hepatosplenomegaly.

The following day, an explorative laparotomy was conducted and a duplication of the proximal part of the jejunum was incidentally discovered (as shown in Fig. 2). (Piperacillin + Tazobactam) 250 mg was injected intravenously immediately, and painting and draping were performed. A transverse incision was made over the right upper quadrant and deepened to the subcutaneous tissue and sheath under general anaesthesia. Upon opening the peritoneum, several findings were noted, including the duplication of the proximal jejunum, an abnormal mesenteric twist and band over the jejunum, a dilated proximal duodenum with a collapsed ileum, and multiple adhesions. Adhesiolysis was performed, and the duplication part of the proximal jejunum was resected (resected specimen shown in Fig. 3) with duodeno-jejuno-anastomosis using Polydioxanone Suture 6.0 R/B. The sheath was closed with Vicryl 4.0 R/B, and the skin was closed with Vicryl 4.0 R/B in the subcuticular fascia. A dressing was applied.

After extubation, the baby was advised to remain Nil Per Oral until further surgery advice. An ultrasonography abdomen showed normal findings, and the baby was then shifted to the nursery ward. The child was kept on IV fluids with N/5 10% dextrose and received potassium chloride injections, Piperacillin + Tazobactam, Amikacin,

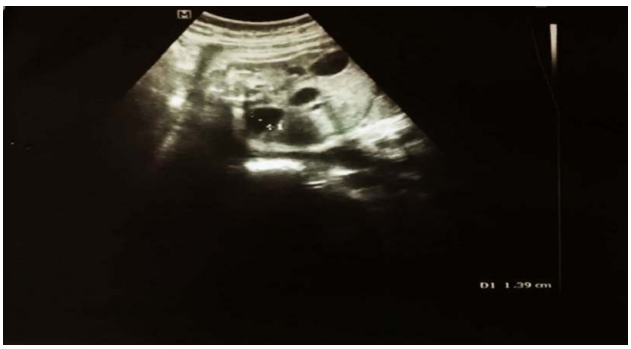

Figure 1. Dilated bowel loops in intrauterine ultrasound measuring up to 13.9 mm seen in the bilateral lower abdominal cavity.

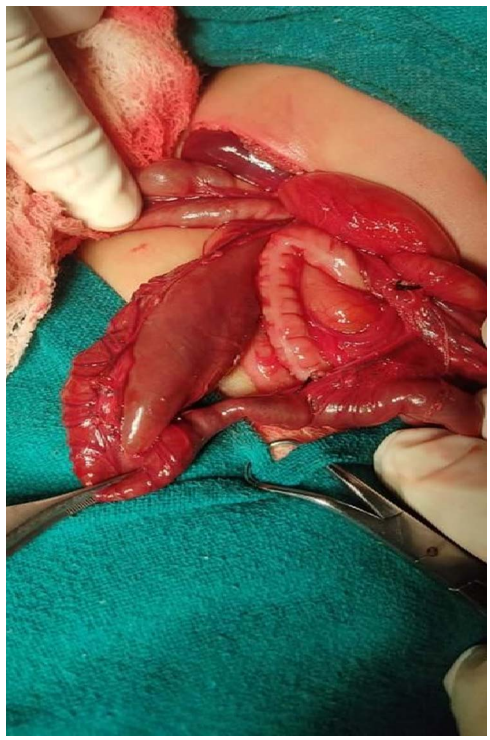

Figure 2. Jejunal duplication cyst discovered as an incidental finding during surgery.

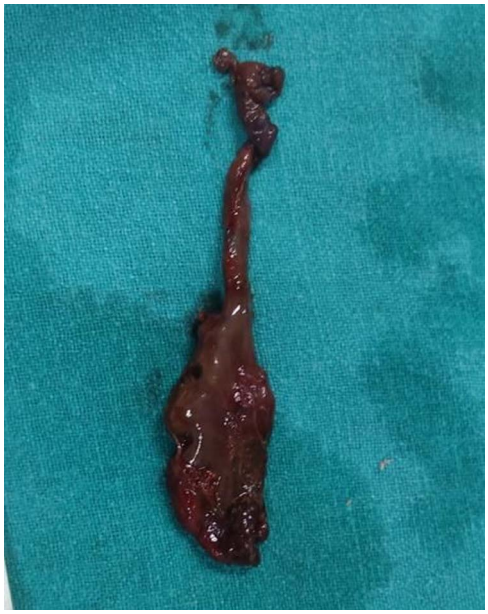

Figure 3. Resected portion of jejunal duplication cyst.

and Fentanyl at the rate of 2 mg QID, Metronidazole, Ranitidine, as well as nebulizations with Adrenaline thrice daily and Budesonide + Hydrocortisone twice daily. The child remained Nil Per Oral for 10 days, during which there was an ongoing issue of decreasing oxygen saturation of the RA. Blood investigations revealed a marked elevation of total leucocyte count (13 200 cells per cubic millilitre) and neutrophilia (79%), for which antibiotics were continued, including injections of (Piperacillin + Tazobactam) and Vancomycin.

The orogastric tube was removed the day after the surgery review, and the child was allowed to start oral feeding while still being kept on antibiotics.

Discussion

Enteric DC are rare congenital malformations that consist of cystic formation in communication with the native gastrointestinal tract, sharing a common muscular wall and blood supply[11]. Their prevalence is noted to be around one in 25 000 deliveries[5]. Duplication of gastrointestinal tract may be located in any part of gastrointestinal system from the oral cavity to the anus[6]. The most common site of enteric DC is the terminal part of the ileum[7]. Duplications are more frequently single. They are usually located in the mesenteric border of the associated native bowel and may vary in shape and size. Most of them are cystic, followed by tubular and mixed type, with or without other congenital anomalies[8]. The aetiology of gastrointestinal duplications has not been well determined, while several theories have been proposed: partial twining, split notochord, environmental factors (trauma or hypoxia), embryonic diverticula, and recanalization defects[12]. It is associated with other abnormalities such as complete colonic duplication, gastric diverticulum, nurenteric cyst and anorectal anomalies[1,2,13]. Ladd described three diagnostic criteria for DC: an intimate attachment to the gastrointestinal tract, a layer of smooth muscle in the wall, and a gastrointestinal epithelial lining[14]. More than 80% of the cases present before the age of 2 years as an acute abdomen or bowel obstruction, but many duplications remain silent unless complications occur, and therefore may not be diagnosed until adulthood and are rarely detected on incidental radiological imaging[8]. In our case, DC of small intestine was suspected in an ultrasound done during routine antenatal visits. Also, the diagnosis was made as an incidental finding intraoperatively. The most common clinical presentations of jejunal DC include signs and symptoms of small bowel obstruction, including vomiting, abdominal pain, bloating, constipation, and abdominal masses[15,16]. However, presentation in our case was increased sleep, decreased feeding, abdominal distension and fever. DC have a smooth muscle layer in the cyst wall, and the cyst is lined with the mucosa of the adjacent alimentary tract, which may contain ectopic digestive tract tissue such as gastric mucosa or pancreatic tissue[17,18]. In our case, the duplication was detected in the jejunum, which is less commonly seen, and was a cystic duplication, the most common type. Cysts with no discernible communication or connection to the adjacent alimentary tract (AT) but with the presence of typical histopathological features of DC would qualify for a diagnosis of an isolated DC[19]. The lining mucosa of DC may not be necessarily that of the adjacent segment of the AT and can be derived from persistent mucosa in the embryonic stage when the duplication occurs. Complications arising from DC are attributed to the presence of heterotopic mucosa in these cysts. Ectopic gastric, pancreatic, squamous, transitional or ciliated mucosa, and ganglion cells can be found in the cyst wall. However, AT duplications are named according to the segment of the bowel in which they occur, rather than the features of their lining mucosa or hamartomatous change[20]. Complications of duplications are numerous like malabsorption, chronic bleeding, mechanical obstruction, volvulus, peritonitis, acute perforation and gastrointestinal bleeding[1,2,4,13,21,22]. Malignant transformation of duplications into adenocarcinoma, neuroendocrine carcinoma, mixed adenocarcinoma, squamous cell carcinoma, leiomyoma and intestinal stromal tumours are well documented[1,2,22]. Mechanical obstruction can be due to external compression from the adjacent duplication, intussusception and volvulus[1,2,21]. Chronic bleeding will present as features of anaemia and melena[13,22]. However, none of abovementioned complications occurred in our patient. endoscopic ultrasound is the diagnostic tool of choice for the investigation of all DC and in some cases, is combined with fine needle aspiration of the cyst to obtain a definitive diagnosis and rule out other more serious pathologies, including potential underlying malignancies[23]. Other diagnostic modalities include plain radiograph of the abdomen, computed tomography, and magnetic resonance imaging, which can all aid in the diagnosis and localization of the cyst[21]. Often jejunal duplications are missed due to its anatomical location. It needs small bowel contrast examination or computer tomography scans to diagnose duplication[1,2,4,21,22]. Surgical treatment is needed in both symptomatic patients and in asymptomatic with a secondary diagnosis for the high prevalence of complications like enteric obstruction, bleeding, volvulus, and rare malignant transformation in adult age[24,25]. The treatment of jejunal DC is mainly surgical and entails complete excision and curation of a primary anastomosis[16]. Same was done in our case as well. As duplication cysts amplify the chances of late malignant transformation, complete excision should be ensured[26]. If the duplication is diagnosed without acute severe complications, excision of the affected jejunal segment with jejunal anastomosis and prophylactic appendectomy (Ladd's procedure)[1,21] or laparoscopic cyst removal[22,27] is the treatment of choice. It will stop the progression of the disease to an acute uncompensated status which will need laparotomy and significant length of bowel resection[1,2,13,21].

Conclusion

We report a case of Proximal jejunal duplication, which is a rare presentation in paediatrics surgery. Surgical intervention is mandatory for this kind of presentation to prevent its varied complications as discussed in discussion part of our case report. It is important to include DC in the differential diagnosis if a neonate presents with features of intestinal obstruction.

Ethical approval

The study is exempt from ethical approval in our institution.

Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

None available.

Author contribution

All the authors were involved in manuscript preparation, review of literature and final approval of manuscript.

Conflicts of interest disclosure

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Research registration unique identifying number (UIN)

Not done (no any new surgical technique or new equipment/ technology used).

Guarantor

Dr. Amrit Bhusal.

Data availability statement

Yes the data analyzed during current study are publicly available, available upon reasonable request, or if data sharing is not applicable to this article.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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