CASE REPORT

Total midgut duplication: a ticking time bomb

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SUMMARY

A day-old neonate presented with bowel obstruction and an abdominal mass. Exploratory laparotomy revealed complete tubular midgut duplication from duodeno-jejunal junction up to terminal ileum which was communicating with the ileum distally. At the proximal end, another 5×5 cm duplication cyst was identified and excised. Postoperatively, complaints were relieved. During follow-up, 99m-Tc-pertechnetate-SPECT scan showed ectopic gastric mucosa in lower abdomen and in the right hemithorax. CECT-chest showed a 3×3 cm foregut duplication cyst, but there were no respiratory symptoms. While being planned for an elective surgery, he presented at 6 months of age in emergency with massive bleed per-rectum and shock. He underwent Wrenn procedure without any injury to the normal bowel. The thoracic foregut duplication cyst was excised later. Total midgut duplication, though benign, may present with life-threatening haemorrhage if left untreated. Presence of one such lesion warrants a search for others. Mucosal stripping is a simple and safe alternate to resection.

BACKGROUND

Total duplications of the small intestine are rare and pose a great surgical challenge. Abrupt haemorrhage with haemodynamic instability due to ulceration, perforation and bleeding can be encountered in duplications with ectopic gastric mucosa. Complete resection is challenging because of the shared blood supply with the adjacent normal bowel and may result in short bowel syndrome. Mucosal stripping and fenestration are other novel alternatives avoiding this complication. A case of total midgut duplication with synchronous foregut duplication cyst having different presentations with age and its successful staged management is described.

CASE PRESENTATION

A 1-day-old neonate presented with bowel obstruction and an abdominal mass. He was born at term by normal vaginal delivery to a 24-year-old primigravida mother. The antenatal period was reportedly uneventful, and no antenatal ultrasound was available. On presentation he weighed 2.5 kg, had tachypnoea, moderate dehydration and cyanosed peripheries. Ultrasound was suggestive of a large mesenteric or an omental cyst. After adequate resuscitation and proper consent, the child underwent exploratory laparotomy. Intraoperatively, a complete tubular midgut duplication extending from the duodeno-jejunal junction up to terminal ileum which was communicating with ileum distally,

was seen ([figure 1A](#)). Another duplication cyst (5×5 cm) was found proximal to the duplicated bowel ([figure 1B](#)). There was associated malrotation of the small bowel. A decision for staged repair was taken because of the poor general condition of the child. The proximal duplication cyst was excised, and the rest of the duplicated bowel was milked and drained distally into the terminal ileum and left as such. The child recovered well postoperatively and was discharged on full oral feeds. The child was kept under close follow-up due to the risk of gastrointestinal (GI) bleeding from rests of potential ectopic gastric mucosa in remainder tubular duplication.

INVESTIGATIONS

The histology of the resected specimen confirmed it to be a duplication cyst. At 5 months of age, he underwent a 99m-Tc-pertechnetate SPECT (single-photon emission computerized tomography) scan, which showed the presence of ectopic gastric mucosa in the lower abdomen and also in the right hemithorax ([figure 2](#)). A chest X-Ray and CECT (contrast enhanced computed tomography) scan revealed a 3×3 cm foregut duplication cyst in posterior mediastinum with obstructive changes seen in the right middle and lower lobes, but there were no respiratory symptoms ([figure 3](#)). While an elective surgery was being planned, he presented a month later in emergency with massive bleeding per-rectum and shock. His haemoglobin had fallen from 11 gm% to 4 gm%. He was stabilised and received multiple blood transfusions.

TREATMENT

After stabilisation, he underwent a second surgery (Wrenn procedure) where the entire mucosal lining of the duplicated bowel was stripped out without injuring the adjacent normal small bowel. The entire mucosal tube measured 90 cm in length and was achieved by giving multiple transverse incisions at regular intervals over the seromuscular layer of the duplicated bowel ([figure 4](#)). The histopathological sections revealed presence of normal gastric mucosa and submucosal tissue.

OUTCOME AND FOLLOW-UP

At 8 months, the child underwent posterolateral right thoracotomy and excision of the foregut duplication cyst. The histology confirmed a foregut duplication cyst with ectopic gastric mucosa and showed areas with extensive haemorrhage and ulcerations. The child had been under regular

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Figure 1 (A) Total midgut duplication. (B) Bowel duplication with proximal portion presenting as cyst.

follow-up until 6 months after the last surgery and was asymptomatic, feeding well and had gained adequate weight.

DISCUSSION

Alimentary tract duplications (ATD) occur in 1 in 18 000 live births and are classically defined by the Ladd's criteria as a congenital lesion that has an anatomical relationship with some part of the bowel, lined by intestinal mucosa and has a well-developed smooth muscle coat.¹ The duplicated bowel could be cystic or tubular and could be present anywhere in the GI tract at the mesenteric side of the normal bowel. The shared common wall and blood supply pose a special surgical challenge in cases of very long tubular duplications.²

The cystic variety is more common, and there is male preponderance.³⁻⁵ Ours is a rare case of long tubular and cystic midgut duplication with associated malrotation and a concurrent cystic foregut duplication cyst.

Clinical presentation varies with the age, size, site, type, number of cysts, communication with bowel and the presence of ectopic gastric mucosa.^{3,4} Antenatal diagnosis is also possible.³ Complications of ATD include obstruction, torsion, rupture, perforation and infection in cases of cystic duplications.⁶ Torrential GI haemorrhage can occur due to ectopic gastric mucosa in duplications, and hence, a complete evaluation using 99mTc-SPECT scan is essential. This child presented just after birth with features of bowel obstruction and underwent cyst excision. The remainder total midgut duplication was left intact after establishing a distal communication with terminal ileum to reduce the operating time in a sick neonate and to avoid any injury to the native bowel as they share a common blood supply. In cases with long tubular duplications, the dilemma regarding complete resection at initial surgery or a staged surgery persists. In sick

Figure 2 (A) Tc-99m-pertechnetate SPECT scan showing ectopic gastric mucosa in lower abdomen and also in the right hemi-thorax. (B) Posterior view showing early tracer uptake by intestinal duplication and delayed uptake in thoracic duplication cyst.

Figure 3 (A) Chest X-Ray showing uniform spherical opacity in the right hemithorax. (B) CECT chest showing compression of right main bronchus. Obstructive changes, crowding of pulmonary vessels in right lung.

neonates, like in ours, a staged surgery is a preferred option as it results in decreased surgical stress, postoperative morbidity and subsequent faster recovery. Despite being under regular follow-up and planned for an elective surgery after initial cyst excision, this patient presented with the dreaded complication of massive GI bleed and shock to the emergency, similar to a few other reported cases.³ After stabilisation, child underwent second surgery in the same hospital admission to prevent any further life-threatening episode of GI haemorrhage.

The treatment of ATD is complete excision. Cystic duplications are easily excised. Problems occur in long tubular duplications like in our case.^{2,6} Excision of the small bowel may result in significant compromise of the bowel length. The alternative surgical options remain mucosal stripping as described by Wrenn or separation of the long tubular duplication segment by dissection of the common wall based on the Bianchi technique.^{2,7,8} In the second surgery, we were able to perform the Wrenn procedure without incurring any injury to the native normal bowel while preserving its blood supply. Sham *et al*² have also reported a similar case where they excised two large cystic duplications of the proximal jejunum and used the Bianchi's principle for the distal tubular duplication segment.

Figure 4 (A) Wrenn's procedure: mucosal tube being created. (B) Mucosal tube about 30 cm in length; multiple transverse incisions on duplicated bowel were given, and 90 cm of mucosal tube was stripped.

In 2%–20% cases, there may be a synchronous thoracic duplication cyst.⁴ Hence, if an ATD is diagnosed, other ATD should be actively searched for like in our case. Thoracic duplications appear to be more common on the right side and have the highest incidence of associated vertebral anomalies, including anterior or posterior spina bifida, hemivertebrae or myelomeningocele.⁹

A simultaneous or a subsequent thoracotomy, according to general condition of the child and the severity of symptoms, is mandatory to eliminate the pathology in cases of

concurrent foregut duplication cyst. This could also be achieved thoroscopically.

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Learning points

- ▶ Alimentary tract duplications may present with intestinal obstruction, abdominal distension, abdominal pain, bleeding and intussusception in neonates.
- ▶ Gastrointestinal bleeding in bowel duplication can be life threatening.
- ▶ Whenever an alimentary tract duplication is encountered actively search for another.
- ▶ Staged surgery in newborns with long duplications can avoid injury to native bowel, but a close follow-up is required in case it bleeds.
- ▶ Mucosal stripping (Wrenn procedure) is a safe and simple alternative to massive bowel resection.

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