

Multiple Enteric Duplication Cysts in a Twin Fetus

Diagnosis and management

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الكيسات المعوية المضاعفة المتعددة في جنين توأم

التشخيص و العلاج

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المخلص: الكيسات المعوية المضاعفة المتعددة هي من التشوهات الخلقية النادرة في الجهاز الهضمي. ويمكن أن يشتهب فيها إذا لوحظت الآفات الكيسية في بطن الجنين أثناء التصوير بالموجات فوق الصوتية قبل الولادة. ويشمل التشخيص التفريقي للآفات الكيسية داخل البطن للجنين على كيسات تربية جنينية، وكيسات مساريقية جنينية، وكيسات العقي الكاذبة وكيسات المبيض الجنيني. نحن نعرض تقرير عن الكيسات المعوية المضاعفة التي تم تشخيصها قبل الولادة في واحدة من الأجنة التوأم والتي تم علاجها بنجاح.

مفتاح الكلمات: التشوهات الخلقية؛ كيسات، حمل توأم؛ التصوير بالموجات فوق الصوتية، التشخيص قبل الولادة؛ ورم صافقي؛ سوء الاستدارة المعوية؛ عائلي، تقرير حالة؛ عمان.

ABSTRACT: Enteric duplication cysts are rare congenital anomalies of the gastrointestinal tract. These can be suspected if cystic lesions are noted in the fetal abdomen during an antenatal ultrasonogram. The differential diagnoses of fetal intra-abdominal cystic lesions include fetal omental cysts, fetal mesenteric cysts, meconium pseudocysts and fetal ovarian cysts. We report an antenatally diagnosed enteric duplication cyst in one of a set of twin fetuses which was managed successfully.

Keywords: Congenital Abnormalities; Cysts; Twin Pregnancy; Ultrasonography; Prenatal Diagnosis; Peritoneal Neoplasm; Intestinal Malrotation, Familial; Case Report; Oman.

ENTERIC DUPLICATION CYSTS ARE RARE congenital anomalies that occur anywhere in the gastrointestinal tract, with a greater predilection towards the ileal region.^{1,2} The reported incidence is 1:4,500 newborns, and the embryogenesis of these anomalies remains controversial.² They may be either cystic or tubular and present at the mesenteric border of the bowel. These can be suspected antenatally if cystic lesions are noted in the fetal abdomen during an anatomy or growth scan. The differential diagnosis of fetal intra-abdominal cystic lesions includes gastrointestinal and urogenital anomalies such as choledochal, hepatic or mesenteric cysts, meconium pseudocysts and fetal ovarian cysts. Prenatal diagnosis of these cystic lesions allows planning for appropriate postnatal work-up and screening for associated malformations.

Only a few cases of antenatally diagnosed duplication cysts are mentioned in English medical literature.³ We report a case of an antenatally diagnosed enteric duplication cyst in one of a set of twins which was managed successfully after birth. A literature review is also included.

Case Report

A 25-year-old gravida 2, para 1, woman with a twin pregnancy presented to Sultan Qaboos University Hospital, Muscat, Oman, in early labour at 35 weeks of gestation. Her antenatal care had been at a private healthcare facility. Her first pregnancy had ended with an intrauterine fetal death at 26 weeks of gestation but had not been investigated further.

The current pregnancy was a spontaneous conception. An early pregnancy scan revealed

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Figure 1: Antenatal scan of fetal abdomen showing two cystic structures.

a dichorionic diamniotic twin pregnancy. Some anomaly was suspected in the second twin during an anatomy scan which would need an evaluation after delivery. A follow-up scan at 29 weeks reported corresponding growth, with the second fetus showing two intra-abdominal cystic structures likely to be related to a duplication cyst and mild polyhydramnios [Figure 1]. The patient had been normotensive and normoglycemic during this pregnancy.

An ultrasound scan, done on admission to the Sultan Qaboos University Hospital, Oman, confirmed a twin pregnancy with the first twin in a cephalic presentation. The second twin, in breech presentation, had two anechoic intra-abdominal cystic lesions that were separate from normal hollow structures (i.e. the bladder and stomach). There was evidence of polyhydramnios.

A decision was made to allow for a trial of vaginal delivery after detailed counselling about the risks to the second twin. The mother progressed to spontaneous vaginal delivery of the first twin but an emergency Caesarean section had to be performed for the second twin due to the transverse lie and prolonged fetal bradycardia. Both neonates were moved to the Neonatal Intensive Care Unit (NICU) for observation and monitoring. The initial assessment of the second twin in the NICU was unremarkable. However, the neonate later started to have persistent green aspirates and an abdominal X-ray showed a dilated stomach, duodenum and proximal small bowel with no gas in the rest of the abdomen, suggesting a small bowel obstruction [Figure 2].

The patient underwent laparotomy soon after stabilisation. A 5 × 7 cm fluid-filled cyst was noted in the mesentery of the proximal jejunum compressing the small bowel lumen,



Figure 2: Plain X-ray of the abdomen showing the gas distended stomach and duodenum but no evidence of gas filled bowel loops distal to it.

causing a complete obstruction [Figure 3]. Further exploration revealed another 6 × 3 cm fluid-filled isolated cyst in the right upper quadrant, extending into the retroperitoneum thereby displacing the duodenum and pushing the extrahepatic biliary system forward. The second cyst also had its blood supply from the mesenteric vessels. The caecum and appendix were positioned in the midline. The cysts were excised completely and Ladd's procedure was carried out to correct the malrotation.

The patient had a smooth postoperative course. An abdominal ultrasound performed before discharge to outline the hepatobiliary anatomy and to rule out any residual retroperitoneal cysts was normal. The histology revealed cyst walls lined by well-formed small bowel type mucosa, submucosa, and circular and longitudinal layers of *muscularis mucosa* confirming small bowel duplication cysts [Figure 4].

Upon the one-year follow-up, the affected child was doing well and achieving normal milestones.

Discussion

Prenatal detection of enteric duplication was first reported by van Dam *et al.* in 1984.⁴ The factors that distinguish enteric duplication cysts from other cystic lesions include thickness of the muscular wall, peristalsis evident in the cystic wall of the enteric duplication and intimate contact of the

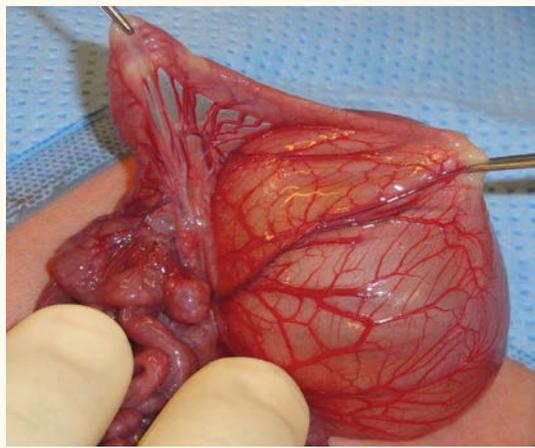


Figure 3: Enteric duplication cyst located at the mesenteric border of the proximal jejunum.

cyst wall with the mesenteric border. Antenatal scans have been reported to identify only 20–30% of such cases, and ours was one of these cases.^{5,6} The neonate in this case was symptomatic for bowel obstruction from the first day of life and a plain X-ray of the abdomen supported the clinical diagnosis.

If the enteric duplication is antenatally undiagnosed and asymptomatic at birth, symptoms usually start during infancy and vary according to the location, size and presence or absence of the heterotopic gastric mucosa. Enteric duplications could act as the lead point for volvulus or intussusceptions.² Gastrointestinal haemorrhage leading to haemodynamic instability, due to ulcerating gastric mucosa within a duplication cyst, has been reported.⁷ Therefore, intestinal duplication cysts should be included in the differential diagnosis for paediatric surgical abdominal emergencies.⁸ Although rare, cases of intestinal carcinomas arising within duplication cysts in adulthood have also been reported.^{9,10} Hence, asymptomatic duplication cysts, when diagnosed incidentally during a surgical procedure, should be excised to prevent future complications as described.^{7,9}

Retroperitoneal enteric duplication cysts are even rarer and most of the reports mention these arising from the left side.¹¹ However, the retroperitoneal cyst in the right upper quadrant in our case causing anterior displacement of the extrahepatic biliary system is, to the best of our knowledge, the first one reported in the literature. Malrotation of the gut, as seen in this case has been reported as a rare associated anomaly with mid-gut

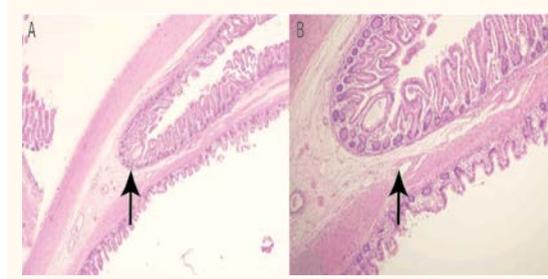


Figure 4 A & B. A: Haematoxylin & eosin stained slide (10 x magnification) showing a duplication cyst in the wall of the intestine; B: Haematoxylin & eosin stained slide (20 x magnification) showing the cyst lined by intestinal type epithelium.

duplication cysts.^{12,13}

A complete resection of the enteric duplication cyst along with the adjacent bowel segment and primary reanastomosis is the recommended treatment for small enteric duplications. If duplication cysts are extensive and involve large segments of the bowel, mucosal stripping is indicated to avoid the risk of short bowel syndrome.^{1,2,12}

Conclusion

Although intestinal duplications are benign lesions in children, they can cause significant morbidity and even mortality if left untreated. With the increasing accuracy of prenatal ultrasounds, intra-abdominal enteric duplication cysts are much more likely to be detected antenatally. Duplication cysts should be considered in the differential diagnosis of abdominal cystic lesions. Surgical excision of the enteric duplications is required to make a definitive diagnosis and to prevent future complications. In our case, prenatal detection of the enteric duplication allowed close neonatal surveillance and timely surgical intervention.

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