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OBSTETRIC CASE REPORTS

# Very early prenatal diagnosis of large isolated thoracic enteric cysts and review of the literature

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## Introduction

Thoracic enteric cysts are extremely rare developmental entities that represent 1–2% of all mediastinal cysts (Davis et al. 1987). In most clinical cases, thoracic enteric cysts are encountered in the late neonatal period or even in childhood, when they are diagnosed because of typical respiratory problems and other symptoms associated with compression of some surrounding structures (Rizalar et al. 1995; Wilkinson et al. 1999; Carachi and Azmy 2002; Altinok et al. 2003; Anagnostou et al. 2009). Other cases are only found incidentally. Prenatal detection of these cystic lesions has rarely been reported in the literature (Newnham et al. 1984; Reisli et al. 2003; Sahinoglu et al. 2005; Cocker et al. 2006). Isolated enteric cysts can migrate to different regions of the body; yet an intra-abdominal location is extremely rare (Sharma et al. 2009), and only four postnatal cases have been described. In this paper, we describe the first very early prenatal diagnosis of large isolated thoracic enteric cysts during an uneventful 1st trimester pregnancy.

## Case report

A healthy 30-year-old pluripara was referred to the Perinatal Unit of the Department of Gynaecology and Obstetrics at 13 weeks' gestation to 'clear up' an ultrasound finding of 'two big holes' in the fetal trunk. Two simple cystic formations were observed by transvaginal sonography. One of the cysts 'filled up' the whole left pleural cavity, compressing the ipsilateral lung tissue and dislocating the heart to the opposite side of the fetal chest. Another transparent cyst with a very thin and smooth wall and without clearly visible vascularisation was situated intra-abdominally on the contralateral side. Both cysts, almost the same size, were identical morphologically and echogenically and were completely filled with fluid. Fetal activity was excellent. At 14 weeks' gestation, a follow-up ultrasound examination revealed significant increases in the size of the thoracic and abdominal cysts, with diameters measuring 30 mm and 34 mm, respectively (Figure 1).

After the local Ethical Committee allowed the pregnancy to be terminated, medical abortion was induced by use of prostaglandins. A dead female fetus of 45 g without external malformations was aborted. Pathological examination confirmed the existence of the thoracic and intra-abdominal cysts, which had transparent and very thin walls and were filled with serous liquid (Figure 1). Both cysts were mobile and the thoracic one compressed the left fetal lung, which was well developed in relation to the gestational age. The cystic formations communicated with each other through a twisted narrowed fibrous canal positioned at the level of the diaphragm, which was not detected by ultrasonography. Based on histopathological features, the cysts were of the simplest type with a single layer of flattened cuboidal serous-type epithelial cells sporadically covering the internal surface of both cysts. There was no trace of intestinal epithelial differentiation. All other thoracic, abdominal and retroperitoneal fetal organs were normal.

## Discussion

Although it is not easy to make an accurate early prenatal diagnosis (Sahinoglu et al. 2005; Eber 2007), the sonographic finding of two transparent cysts with very thin and smooth walls, localised in the fetal thorax and abdomen, was very suggestive of thoracic enteric cysts. Enteric cysts are classified as mediastinal developmental lesions that are thought to be identical to neurenteric cysts, but without communication with the vertebral column or spinal canal (Sharma et al. 2009). Most likely, they are part of a spectrum of the same embryonic disorder, rather than separate

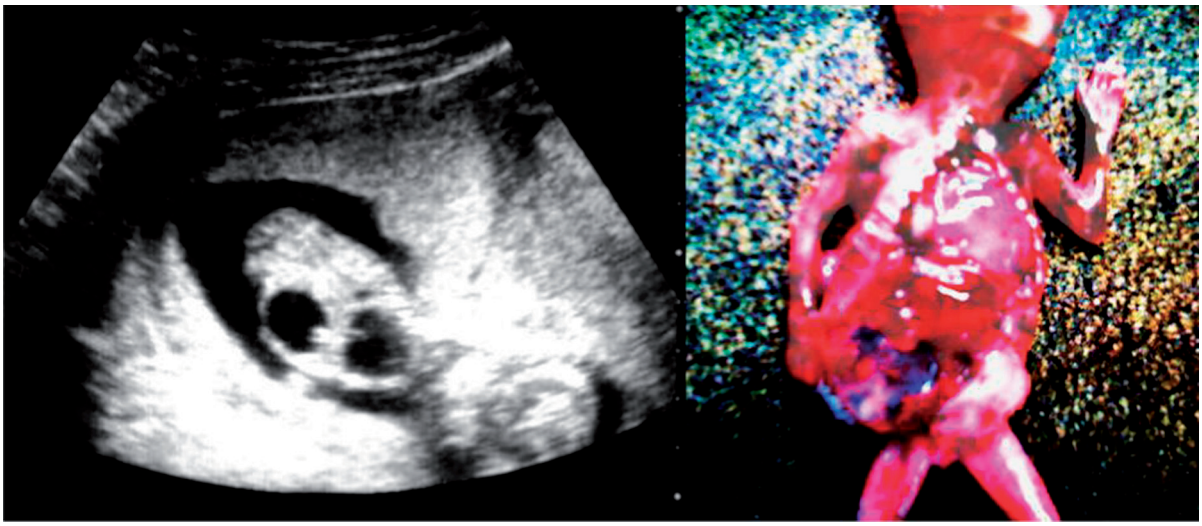

Figure 1. Thoracic enteric cysts located in the fetal left pleural cavity and right hemi-abdomen: sonographic scan at 14 weeks' gestation and macroscopic appearance after termination of the pregnancy.

Table I. Prenatally detected cases of thoracic enteric/neurenteric cysts (in chronological order).

| Authors | Type of cysts | GA (weeks) | Morphology, size of cysts | Associated abnormalities | Location | Communication | Therapy | Outcome |
|---|---|---|---|---|---|---|---|---|
| Newnham et al. (1984) | Enteric | 34 | Single, multilocular, clear, 63 × 34 mm | No | PM, right HT | No | Thoracotomy | VBT, good |
| Rizalar et al. (1995) | Neurenteric | 32 | Single, unilocular, clear, 40 × 50 mm | Scoliosis, SB, hemivertebrae | PM, right HT | No | Thoracotomy | CST, good |
| Perera and Milne (1997) | Neurenteric | 18 | Single, unilocular, clear, 46 × 24 mm | Hemivertebra, defect at Th3–4 | PM, right HT | No | Thoracotomy | VBT, good |
| Olavarria et al. (2000) | Neurenteric | 35 | Single, unilocular, transparent, large | Lung hypoplasia, hemivertebra, macrocephalia | PM | CNS | Not applicable | VBT, died |
| Wilkinson et al. (1999) | Neurenteric | 28 | Single, bilocular, transparent, 50 mm | Hemivertebrae Th1–3, fetal hydrops | PM, right HT | No | Prenatal TA, thoracocentesis, thoracotomy | PVB, good |
| Uludag et al. (2001) | Neurenteric | 34 | Single, unilocular, clear, 31 × 44 mm | Scoliosis, hemivertebrae, pyelectasia bilateral | PM, right HT | No | Thoracotomy | PVB, good |
| Reisli et al. (2003) | Enteric | 32 | Single, unilocular, transparent, large | Pectus excavatum, compression of thoracic duct | PM, right HT | No | Prenatal aspiration, thoracotomy | Good |
| Sahinoglu et al. (2005) | Enteric | 22 | Single, unilocular, clear, 50 × 27 mm | No | PM, right HT | No | Thoracotomy | VBT, good |
| Cocker et al. (2006) | Enteric | ? | Multiple, unilocular, tubular, large – 5 mm | No | Right HT, left hemi-abdomen | No | Thoracoscopic/laparoscopic resection | VBT, good |
| Bernasconi et al. (2007) | Neurenteric | 38 | Double, unilocular, clear, 20 mm and 40 mm | Thoracic vertebral defect | PM, right HT | Spinal canal | Thoracotomy | CST, good |
| Kimya et al. (2007) | Neurenteric | 21 | Single, multilocular, seromucinous, 25 × 11 mm | Vertebral defect at Th5–7 | PM, right HT | Epidural space | Not applicable | Induced abortion |
| Aydin et al. (2009) | Neurenteric | 28 | Single, unilocular, mucinous, 26 × 15 mm | Scoliosis, cleft vertebra C7 | PM, cervical | Spinal canal | Postnatal resection | CST, good |
| Gadodia et al. (2010) | Neurenteric | 28 | Double, unilocular, clear, 40 × 50 mm | Vertebral segmentation | PM | Spinal canal | Thoracotomy | VBT, good |
| Present case | Enteric | 13 | Double, unilocular, clear, 30 mm and 34 mm | No | Left HT, right hemi-abdomen | Between the cysts | Not applicable | Induced abortion |

GA, gestational age; PM, posterior mediastinum; HT, hemithorax; VBT, vaginal birth at term; SB, spina bifida; TA, thoracoamniotic shunt; PVB, premature vaginal birth; CST, caesarean section at term

entities. Detachment of the abnormal bud from the foregut, determined for gastrointestinal differentiation, should result in the development of enteric cysts. However, neurenteric cystic lesions of endodermal origin with a vertebral dysplasia usually develop as a consequence of failed complete separation of the notochord from the foregut (split notochord syndrome) during the 3rd postconceptional week (Almog et al. 2001; Reisli et al. 2003; Sharma et al. 2009; Aydin et al. 2009). According to different theories, the developing foregut adheres focally to the notochord, leading to growth of a traction diverticulum and resulting in a foregut cyst and concomitant vertebral anomalies (Veeneklass 1952). Another theory advocates the gut herniates into a gap formed by a split notochord (Almog et al. 2001). In about 40–70% of cases, these cysts can be accompanied by other congenital abnormalities such as hemivertebrae, scoliosis and spina bifida (Uludag et al. 2001; Carachi and Azmy 2002; Altinok et al. 2003; Reisli et al. 2003; Setty et al. 2005; Gadodia et al. 2010). In one-third of the patients, these cysts are associated with CNS and/or gastrointestinal tract malformations, while intracranial neurenteric cysts are very uncommon.

Thoracic enteric cysts can occur in both males and females (Carachi and Azmy 2002) and occur at a very low incidence; the combined existence of two cysts at opposite sides of the diaphragm is particularly rare (Altinok et al. 2003; Cocker et al. 2006). Approximately 20 fetuses with anomalies are seen yearly at our department, but this is our first prenatal detection of thoracic enteric cysts in a 25-year period. Previously detected and reported cases with a prenatal diagnosis of thoracic enteric/neurenteric cysts are presented and compared with our case in Table I (Perera and Milne 1997; Olavarria et al. 2000; Bernasconi et al. 2007; Kimya et al. 2007). These reports did not allow any conclusion about early development of thoracic enteric cysts. It can only be summarised that these cysts have been commonly detected in the 3rd trimester of pregnancy, and mostly are single, unilocular, transparent and located in posterior mediastinum and right hemithorax. In contrast to the reported cases, we would like to emphasise that the findings in the present case demonstrate that these cysts developed in early embryogenesis. The very thin and smooth walls of the two cysts, transparent serous contents, internal layer of low cuboidal epithelium due to intracystic pressure, and the physical junction by means of a connective tissue canal could represent histopathological characteristics of thoracic enteric cysts occurring at an early phase of embryological development. Most likely, later during development, the thin-walled cysts would become thicker and the fluid content would change according to the maturation process of the epithelial cells, which become more differentiated and functional. We can only speculate that a definitive separation of the two cysts may be the result of accelerated fetal growth and apoptotic processes may be responsible for the obliteration. The autopsy confirmed an evident transdiaphragmatic connection between the two lesions, which could have been the developmental base for the muscle layer of the primitive intestine and may have been obliterated after further development. This connection between the two cystic structures undoubtedly indicates their common embryonic origin. Early in embryonic life, the thoracic and abdominal cavities are connected to the pericardioperitoneal canal, which is reflected by the communication between the cysts.

The differential diagnosis of thoracic enteric cysts includes: thoracic duct cysts; bronchogenic cysts; congenital cystic adenomatoid malformation; oesophageal duplication cysts; neurenteric cysts; diaphragmatic and hiatal hernias; gastrointestinal duplication cysts; pericardial and paracardial cysts; dermoid cysts; cystic hygroma and hydatid cysts (Davis et al. 1987; Altinok et al. 2003; Wilkinson et al. 1999; Bernasconi et al. 2007; Traibi et al. 2010). Their natural histories are extremely variable. These cystic lesions can grow, but many of them decrease in size before birth, and some are no longer detectable in the neonatal period. For this reason, some authors do not recommend aspiration of the cyst *in utero* as a definitive treatment (Wilkinson et al. 1999; Reisli et al. 2003; Eber, 2007). Antenatal intervention is only required if fetal hydrops develops (Wilkinson et al. 1999; Sahinoglu et al. 2005; Bush, 2009). Management in the postnatal period is also controversial. Namely, some authors suggest simple long-term observation; while, most authors advocate elective resection of all cystic malformations, especially if they are symptomatic (Eber, 2007). While imaging methods are suggested for the diagnostic evaluation, a histopathological finding after surgical resection of the cyst is usually required for an accurate postnatal diagnosis (Newnham et al. 1984; Altinok et al. 2003).

To our knowledge, this is the first case of an early prenatal diagnosis of isolated thoracic enteric cysts, with very clear evidence of a direct connection between the cysts, as well as the pleural and peritoneal cavities, at the early stages of embryological development. Based on the different morphologies and histologies between postnatally and prenatally detected cases, we can only speculate that these cysts would have had great developmental changes during fetal life. A possible clinical solution for the very early prenatal diagnosis of large thoracic enteric cysts, which could result in lung hypoplasia, fetal hydrops and have poor prognosis, is to allow the termination of such a pregnancy. However, evolution of the cystic lesions could also be followed at any time before the abortion time limit, while waiting for a definitive decision, because the vital prognosis is different in cases of small isolated cysts that can be managed conservatively. Considering the small number of cases worldwide and the great spectrum of congenital thoracic abnormalities, qualitative antenatal and postnatal counselling of the family should be on a case-by-case basis (Bush, 2009). All the reported cases of this very rare abnormality are helpful in clarifying the aetiology, incidence and relationship between the initial phases of embryological development and the developed tumour-like cystic masses.

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