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Letters to the Editor

Prenatal diagnosis of the rarely observed split notochord syndrome

Split notochord syndrome (SNS) is a very rare, occult spinal dysraphism syndrome that can present within a wide spectrum of malformations. SNS can be as mild as a neuroenteric cyst, or as severe as a dorsal enteric fistula, accompanying severe intracranial malformations and malformations of the nervous system (e.g. diastematomyelia, diplomyelia, myelomeningocele)[1]. We report a case of a fetal thoracic cystic mass, observed and suspected to be SNS prenatally and confirmed as such postnatally. To our knowledge, this is the fourth prenatally detected case to be reported in the literature.

A 35-year-old primigravida was referred to our department at 21 weeks of gestation with a suspicious intrathoracic cystic mass seen on fetal ultrasound examination. Detailed ultrasound examination revealed a 25 × 11-mm posterior thoracic hypoechoic bilobar cystic lesion extending towards the right basal pulmonary region (Figure 1a). The neighboring vertebral bodies were deformed and cleaved (Figures 1b and 2). No remarkable findings were observed in other anatomical planes. Amniotic fluid volume was normal. SNS was diagnosed putatively on the basis of the ultrasound findings. However, the presence of an anterior meningomyelocele, which ultrasound examination should reveal as being located more anterolaterally than SNS, could not be ruled out[2]. Differential diagnoses of a cystic mass at this location also include diaphragmatic Morgagni hernia, esophageal duplication, congenital cystic adenomatoid malformation type I, neuroblastoma, thymic cyst, bronchogenic cyst, pulmonary sequestration or neuroblastoma[3]. Amniocentesis and fluorescence *in-situ* hybridization analysis revealed a normal 46,XY karyotype. The case was discussed by the perinatal ethics committee. Taking into account the possible grave sequelae that this malformation would pose even if the fetus could survive, elective termination of the pregnancy was offered to the couple at 24 weeks of gestation.

The couple gave permission for the pathological examination of the fetus following termination. Macroscopic examination of the abortus revealed a 3 × 2 × 1.5-cm seromucinous multiloculated cyst, located in the posterior mediastinum between the heart and the posterior thoracic wall, which was in communication with the epidural space through a midline defect in the fifth, sixth and seventh vertebral bodies (Figure 3). The cyst wall was demarcated from the dura mater and extended into the medullary space. There was no sign of posterior dysraphism in the neural tube. Thoracic and abdominal organs were macroscopically normal. Histopathological examination revealed a multiloculated cystic formation with a fibrous wall containing smooth muscle fibers. The lumen was lined with a single layer of partially pseudostratified, putatively primitive enteric columnar epithelium with

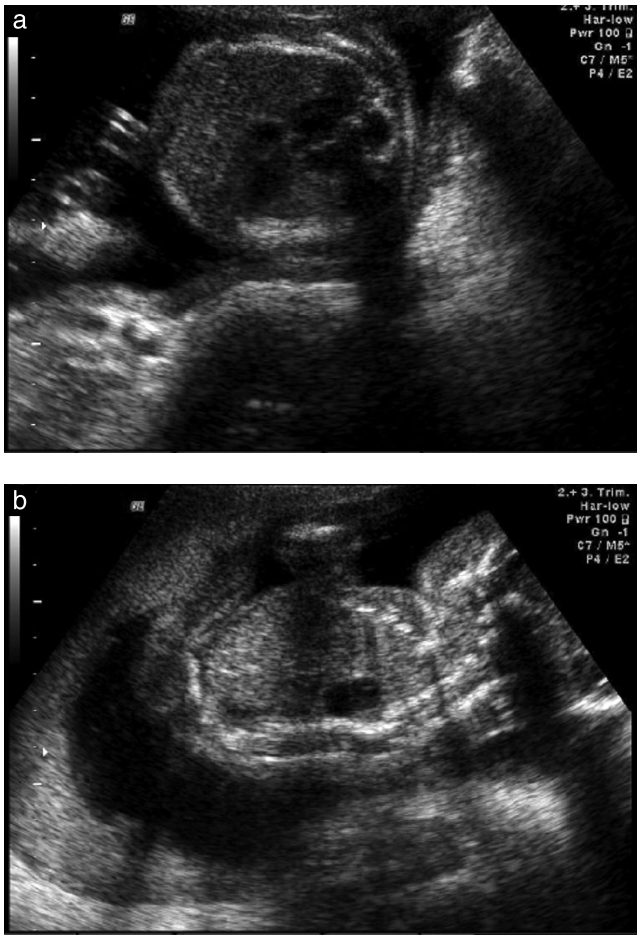

Figure 1 Transverse (a) and sagittal (b) fetal ultrasound images at 21 weeks' gestation showing a posterior thoracic cystic mass.

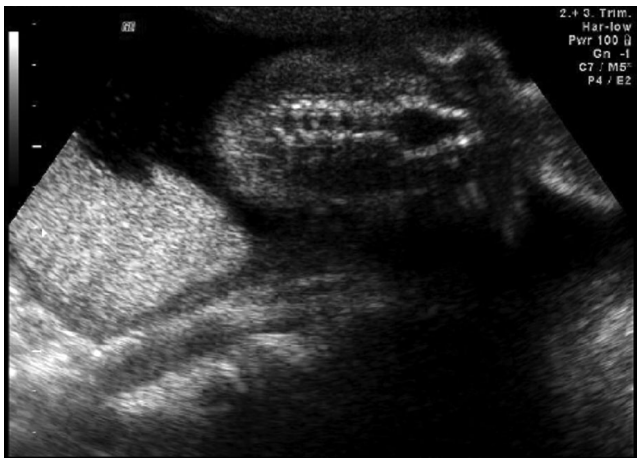

Figure 2 Coronal ultrasound image showing the vertebral defect that was associated with the intrathoracic mass.

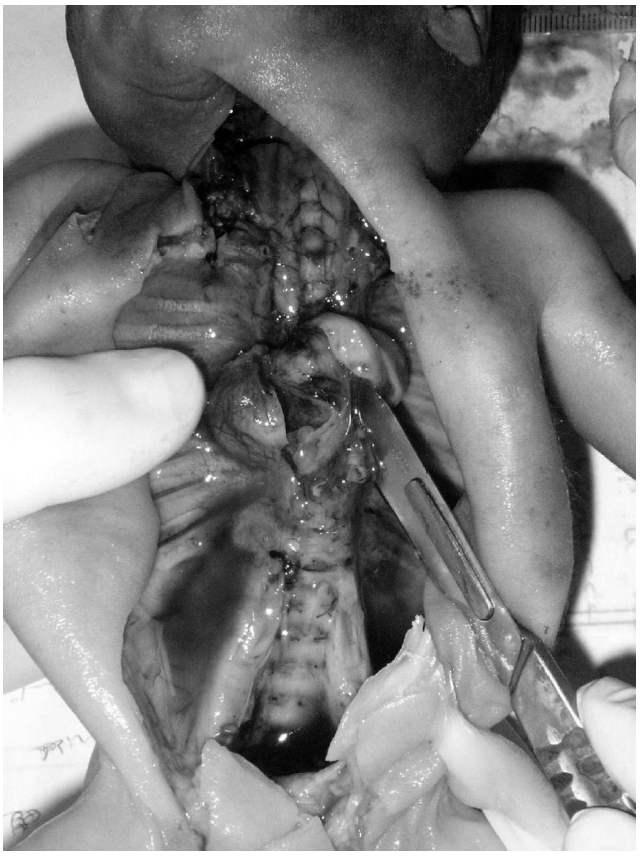

Figure 3 Macroscopic view of the cystic mass during pathological sectioning.

basally located nuclei. Immunohistochemical examination of the cyst wall showed positive staining with cytokeratin 7 (CK7) antibodies and negative staining with CK20. Smooth muscle actin confirmed the presence of smooth muscle fibers, leukocyte common antigen the lymphoid aggregates, and S-100 the peripheral nerve fibers. There was no positive staining for glial fibrillary acidic protein in the cyst wall, ruling out the presence of a neural component to the cyst.

There are only three prenatally diagnosed cases of SNS reported in the literature (two by Almog *et al.*[3] and one by Agangi *et al.*[4]). In two of these SNS with thoracic openings is described, whereas in the third SNS in the sacrococcygeal region is reported[3,4]. In one of these cases, SNS was diagnosed just following delivery, whereas in the other two it was diagnosed at 25 and 31 weeks of gestation. Two cases had polyhydramnios. One case delivered following preterm premature rupture of membranes; the other two delivered at term. Two cases were followed up by neurosurgery and pediatric surgery. Four years postnatally, one case, reported by Almog *et al.*, underwent relocation of the stomach to below the diaphragm from its previous location in the posterior mediastinum. The other case, reported by Agangi *et al.*, underwent an initial colostomy followed by anorectoplasty, an anorectal fistula operation and a lipomyelomeningocele operation to help improvement of the neurogenic bladder and bowel function. Both cases reported by Almog *et al.* were severely kyphoscoliotic, with one exhibiting severe developmental delay most probably due to feeding problems. In the case reported by Agangi *et al.*, neurogenic bladder and bowel dysfunction persisted despite intervention. Unlikely differential diagnoses in this case were a diaphragmatic hernia, usually visualized in the left hemithorax, a thymic cyst, usually located in the anterior mediastinum, and a bronchogenic or pulmonary abnormality, not usually associated with vertebral malformations. The vertebral deformity was far larger than that of a vertebral 'dissipation' potentially caused by a neuroblastoma.

The prognosis of SNS cases diagnosed postnatally has been revised and reported to be poor by Akgur *et al.*, with 9/14 cases undergoing termination, four surviving and one being lost to follow-up[5]. It therefore seemed reasonable to offer termination of pregnancy to the couple as a complex malformation possibly involving the neural axis pointed to a probable diagnosis of SNS. The prenatal identification of a thoracic cyst necessitates ruling out the presence of any associated defect in the vertebral column and the gastrointestinal tract. A neuroenteric cyst or SNS should be considered among other possible pathologies.

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Two- and three-dimensional transvaginal Doppler ultrasound analysis of urethral clear cell carcinoma

A 78-year-old woman, gravida 6 para 3, had suffered from urinary difficulty for more than 5 months. She presented at our clinic complaining of having had vaginal discharge with a foul odor for 1 month. Pelvic