Prenatal Diagnosis of Pulmonary Sequestration by Ultrasound and Magnetic Resonance Imaging

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A 36-year-old multigravida, G2P1, underwent routine ultrasound scan at 22^{+1} weeks of gestation, which revealed a single normally growing fetus with left intrathoracic mass and left displacement of the cardiac apex. The left intrathoracic wedge-shaped hyperechogenic mass, measuring 32 × 25 mm in size, was situated at the lower portion of the left lung. A combination of color and power Doppler ultrasound allowed visualization of a vessel arising from the descending aorta, which supplied the mass. The diagnosis of extralobar pulmonary sequestration was made. Magnetic resonance imaging (MRI) was also performed and revealed a well-defined mass with homogeneous high-signal intensity when compared with normal lung tissue in the left upper lung field, which was compatible with pulmonary sequestration. The pulmonary mass was followed up by color and power Doppler every 2 weeks. The peak velocity of 11.85 cm/sec and the diameter of the feeding artery of 1.19 mm gradually decreased and disappeared 8 weeks later. The intrathoracic mass disappeared 10 weeks later at 32^{+1} gestational weeks. Repeat MRI also revealed spontaneous regression of the mass in favor of resorption of sequestration. The fetus was delivered at 38^{+1} gestational weeks. A male newborn weighing 2,520 g was spontaneously delivered with an Apgar score of 8 at 1 minute and 9 at 5 minutes. In our patient, it is suggested that progressive decreases in the peak velocity of the feeding vessel heralded the spontaneous regression of pulmonary sequestration not associated with hydrops/hydrothorax. [*J Chin Med Assoc* 2008;71(1):53–57]

Key Words: magnetic resonance imaging, pulmonary sequestration, ultrasound

Introduction

Pulmonary sequestration is a congenital anomaly consisting of an abnormal mass of lung tissue which does not communicate with the tracheobronchial tree and is supplied by a systemic artery. Prenatal diagnosis of the intrathoracic type has been reported,[1] but differential diagnosis with other thoracic space-occupying lesions may be difficult.[2–4] Demonstration of the anomalous vascular connection has been achieved postnatally by using angiography and color Doppler investigation and prenatally by using Doppler ultrasound.[1,2] Here, we report a case of pulmonary sequestration that was diagnosed prenatally on the demonstration of anomalous vascular connection using color Doppler ultrasound. The lesion spontaneously regressed *in utero* 8 weeks later. We also review the literature on spontaneous regression of pulmonary sequestration.

Case Report

A 36-year-old multigravida, G2P1, underwent routine ultrasound scan at 22^{+1} weeks of gestation, which revealed a single normally growing fetus with a left intrathoracic mass and left displacement of the cardiac apex. The left hyperechogenic intrathoracic mass was wedge-shaped, measured 32 × 25 mm in size, and was situated at the lower portion of the left lung (Figure 1A). The diaphragm appeared intact, and no pleural effusion, hydrothorax or polyhydramnios was seen. A combination of color and power Doppler ultrasound

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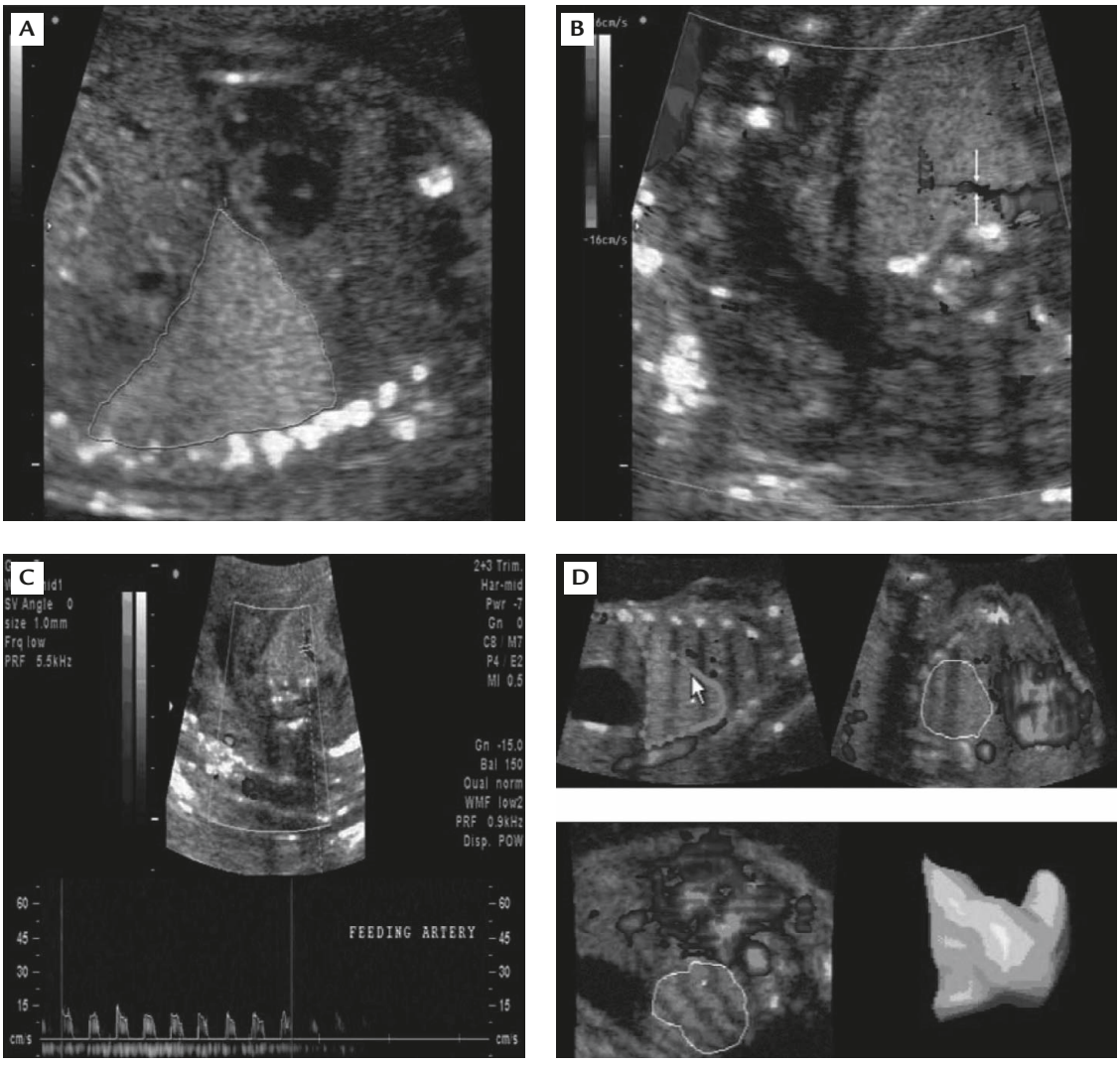

Figure 1. (A) Prenatal 2-dimensional ultrasound study of the fetal thorax in longitudinal section. The pulmonary sequestration is shown as high echogenic intensity at the left lower lobe of the fetal lung at 22^{+1} gestational weeks. (B) Color Doppler ultrasound of the fetal thorax in transverse section. The feeding vessel originates from the descending aorta. The diameter of the feeding artery is 1.19 mm. (C) Color Doppler ultrasound of the fetal thorax in transverse section. The peak velocity of the feeding artery is 11.85 cm/sec. (D) Three-dimensional measurement of the size of the pulmonary sequestration was 3.08 cm^3.

allowed visualization of a vessel arising from the descending aorta, which supplied the mass (Figures 1B and 1C). A volume of 3.08 cm^3 of the intrathoracic mass was measured by 3-dimensional ultrasound (Figure 1D). The diagnosis of extralobar pulmonary sequestration was made. The karyotype of the chromosome was normal (46,XY).

Fetal echocardiography demonstrated displacement of the heart, with normal anatomic structure and vascular connections. Subsequently, magnetic resonance imaging (MRI) was performed by a 1.5T MR scanner (Twin Excite; GE, Milwaukee, WI, USA) with 8-channel phase-arrayed body coils. The mother's pelvis was surveyed in transaxial and coronal view by single-shot fast spin-echo (SSFSE; GE). The presence of a well-defined mass with homogeneous high-signal intensity when compared with normal lung tissue in the left lower lung field was compatible with pulmonary sequestration (Figure 2).

After consultation with the ethics committee of the hospital, conservative treatment for the fetus was recommended. The pulmonary mass was followed-up by color and power Doppler ultrasound every 2 weeks. The diameter of the feeding artery started out at 1.18 mm, gradually decreased and eventually disappeared 8 weeks later. The mass and its feeding artery disappeared completely 10 weeks later at 32^{+1} gestational weeks. Repeat MRI revealed spontaneous

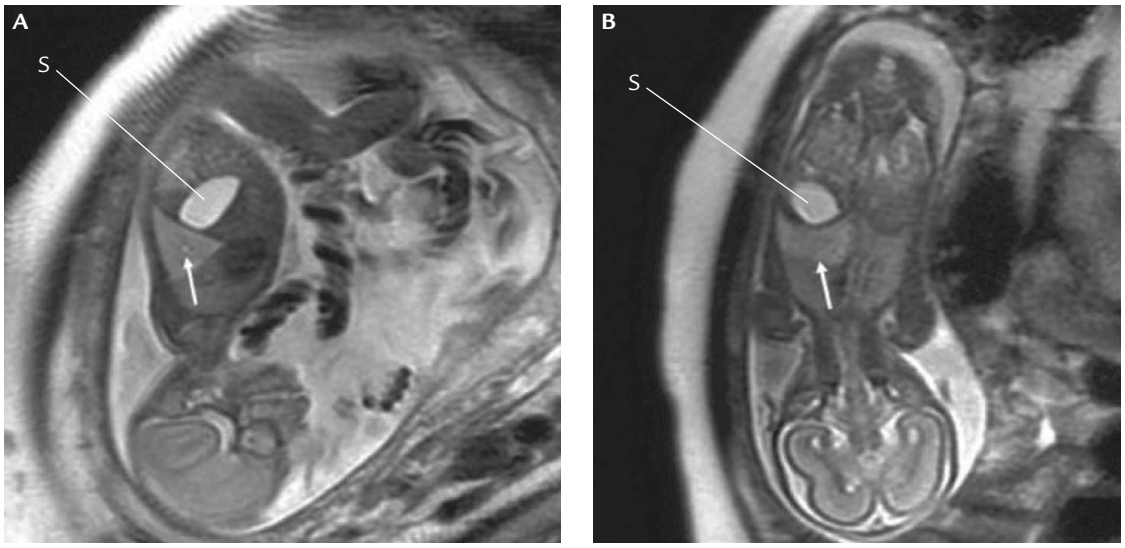

Figure 2. Magnetic resonance images of the pulmonary sequestration. (A) Prenatal magnetic resonance study in sagittal section of the pulmonary sequestration at 22^{+1} gestational weeks. (B) Coronal section of the pulmonary sequestration in the fetal lung. S = stomach.

regression of the mass. The previously identified sequestration lesion could not be identified in this study, favorably suggesting resorption of sequestration.

The fetus was delivered at 38^{+1} gestational weeks. The male newborn, weighing 2,520 g (at the 5th percentile of normal reference weight), was spontaneously delivered with Apgar scores of 8 at 1 minute and 9 at 5 minutes. The pulmonary function of the newborn was checked and revealed oxygen saturation of 98% at room air. There were no other signs of pulmonary anomalies.

Discussion

Pulmonary sequestration is a mass of pulmonary tissue that lacks communication with the bronchial tree, with blood supply arising from an anomalous systemic vessel commonly originating from the descending aorta. The appearance of pulmonary sequestration on prenatal ultrasound examination is similar to that of microcystic congenital cystic adenomatoid malformation of the lung.[5] Mixed or hybrid tumors containing both pulmonary sequestration and congenital cystic adenomatoid malformation are common. In the absence of hydrops, pulmonary sequestration has a favorable outlook which justifies watchful waiting.[5] Many cases show spontaneous regression during the third trimester.[5] Microcystic congenital cystic adenomatoid malformation and pulmonary sequestration with hydrops have an almost 100% perinatal mortality rate.[6,7]

Pulmonary sequestration has been shown to frequently coexist with congenital cystic adenomatoid malformation, and a common etiology has been suspected.[8] Identification of a systemic artery (a characteristic of pulmonary sequestration) is critical in evaluating any spontaneous intrauterine regression. Color Doppler ultrasound imaging is able to identify these arteries, but 3-dimensional power Doppler ultrasonography may be even more sensitive.[9] Progressive hydrops during the second trimester carries such a poor prognosis that prenatal intervention should be considered. A solid tumor with its own blood supply may be treated with a laser. Interstitial laser coagulation has been described for fetal sacrococcygeal teratoma, twin reversed arterial perfusion sequence and chorioangioma.[10] Bruner et al described a failed attempt at using a laser to reduce tumor size in a fetus with microcystic congenital cystic adenomatoid malformation.[10] Davenport et al reported 67 cases of fetal lung lesions, including 1 fetus with pulmonary sequestration that was treated with a laser, but the fetus died postnatally.[7]

The pathophysiology of hydrothorax associated with sequestration is not completely understood. It has been suggested that it could result from torsion of the mass around its vascular pedicle,[11] but this is not likely in some cases since the pedicle was identified by color Doppler. In cases in which the mechanism of hydrops involves high-output cardiac failure[12] in addition to mediastinal compression,[13] other prenatal therapeutic strategies such as alcohol ablation of the pedicle or medical treatment of heart failure may be indicated.

The cause of ipsilateral effusion associated with pulmonary sequestration remains uncertain. It has been postulated that the vascular pedicle leading to the

Table 1. Review of 23 prenatally diagnosed pulmonary sequestrations associated with pleural effusion or hydrops

Reference	Year	Gestation at diagnosis (wk)	Pleural effusion	Hydrops	Prenatal intervention	Outcome
Chan et al[14]	1996	30	Yes	No	Shunt*/sclerosis	Survival
da Silva et al[14]	1996	34	Yes	Yes	None	Survival
da Silva et al[14]	1996	30	Yes	Yes	None	Survival
da Silva et al[14]	1996	25	Unknown	Yes	None	Survival
Evans[14]	1996	30	Yes	Yes	None	Survival
Evans[14]	1996	25	Unknown	Yes	None	Survival
Evans[14]	1996	34	Unknown	Yes	None	Survival
Cass et al[14]	1997	19	Unknown	Yes	Fetal resection	Survival
Cass et al[14]	1997	32	Yes	No	None	Death
Becmeur et al[14]	1998	20	Unknown	Yes	Paracentesis	Survival
Becmeur[14]	1998	30	Yes	Yes	Shunt*	Survival
Weist & Raudies[14]	1999	30	Yes	No	Serial thoracentesis	Survival
Lopoo et al[14]	1998	20	Yes	No	Thoracentesis/shunt*	Survival
Lopoo et al[14]	1998	30	Yes	Yes	Shunt*	Survival
Salomon et al[16]	2003	34	Yes	Yes	Thoracoamniotic/shunting	Survival
Nicolini et al[2]	2000	26	Yes	Yes	Alcohol injection	Survival
Ruano et al[17]	2005	20	No	No	Embolization	Survival
Ruano et al[17]	2005	17	No	No	Embolization	Survival
Ruano et al[17]	2005	31	No	No	Open lobectomy	Survival
Oepkes et al[5]	2007	21	Yes	Yes	Laser treatment	Survival
Harmath et al[18]	2007	21	No	No	No	Terminated
Harmath et al[18]	2007	21	No	No	No	Live born but died of abrupt placenta
This patient	2008	22	No	No	No	Survival

**Thoracoamniotic shunt.*

sequestration can become twisted, resulting in venous and lymphatic obstruction.[14] Alternatively, effusion may develop from a large pressure gradient between a systemic artery and the pulmonary vein.[14] Regardless of etiology, large persistent effusions cause not only lung compression leading to pulmonary hypoplasia but can also result in possible vena caval compromise due to mediastinal shift. This cascade of pathophysiology leads to hydrops and the associated risk of premature delivery and perinatal death. Draining these effusions *in utero*, thereby decompressing the fetal thorax, can improve survival.[15] However, the means to effect adequate drainage remain controversial. Some authors advocate serial thoracentesis, whereas others recommend thoracoamniotic shunting.[15] There are also reports of successful expectant management in hydropic fetuses with pulmonary sequestration. Thoracocentesis alone often results in rapid re-accumulation of fluid, presumably because the underlying pathophysiology leading to the effusion has not been corrected. It may be more logical, therefore, to place a thoracoamniotic shunt to decompress the fetal thorax.[15]

The outcomes of 23 pulmonary sequestrations were reviewed (Table 1).[2,5,14,16–18] Twelve (52.1%) of them were associated with hydrops, and 8 of the 12 fetuses (66.6%) survived without prenatal intervention. Four (4/23, 17.3%) were associated with no hydrops and all 4 survived without prenatal intervention. The review strongly suggests that for pulmonary sequestration associated with no hydrops, no prenatal intervention is needed as it will most likely regress spontaneously.

At present, ultrasound and MRI can reliably differentiate pulmonary sequestration from other lung lesions. This distinction allows accurate prenatal counseling, surveillance, and formulation of a treatment plan based on the known relatively favorable natural history of pulmonary sequestration. The mechanism behind regression of pulmonary sequestration remains unclear, but it is probably the result of vessel constriction of the parenchyma small arteriole resulting in decreasing peak velocity of the feeding artery. This in turn obliterates the vessel, with simultaneous absence of the flow of the feeding artery, as shown in our case. Furthermore, it is suggested that progressive decline of the peak velocity of the feeding vessel can herald spontaneous regression of the pulmonary sequestration not associated with hydrops/hydrothorax. Conservative

observation could be the best option for pulmonary sequestration not associated with hydrops or pleural effusion in the fetal lung and when the peak velocity of the feeding vessel has shown progressive decline.

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