Taiwanese Journal of Obstetrics & Gynecology 56 (2017) 402–405

Contents lists available at ScienceDirect

Taiwanese Journal of Obstetrics & Gynecology

journal homepage: www.tjog-online.com

Case Report

Complete resolution of hydrops by placement of double basket catheter in a case of macrocystic type multilocular pulmonary sequestration☆

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ARTICLE INFO

Article history:
Accepted 21 March 2017

Keywords:
Congenital lung cyst
Macrocystic lung lesions
Hydrops fetalis
Prenatal intervention
Thoracoamniotic shunts

ABSTRACT

Objective: We presented a fetus affected by macrocystic lung lesions with progressive hydropic changes during the second trimester, but experienced remarkable resolution of hydrops in the third trimester after a series of *in utero* interventions.
Case report: A 19-year-old women, G1P0, presented with fetal multilocular thoracic mass and hydropic change at 23^{+4} weeks of gestation. After non-directive genetic counseling, she opted for intrauterine cyst aspiration followed by intra-cystic OK-432 injection at 24 weeks of pregnancy, as well as sequential thoracoamniotic shunts at 26 weeks and 27^{+3} weeks of pregnancy when we observed hydrops developed progressively. Finally, the hydrops resolved in the third trimester and a healthy baby was born at 33^{+3} weeks of pregnancy, in which further surgical intervention was performed at five-month old.
Conclusion: Thoracoamniotic shunting is a preferred option for all hydropic fetuses resulted from large macrocystic lung lesions to enhance perinatal survival rate.

Introduction

Congenital pulmonary cyst is a rare anomaly sometimes leading to stillbirth if hydrops fetalis developed. The differential diagnoses include congenital cystic adenomatoid malformations (CCAMs, also refer to congenital pulmonary airway malformation (CPAM)), bronchopulmonary sequestration (BPS), congenital lobar emphysema, and mediastinal teratoma [1]. In fact, a more practical classification was proposed only using either microcystic (*i.e.* solid appearance on fetal ultrasound) or macrocystic type [2], because it was considered that the prognosis of fetuses with macrocystic lung lesion turned out to be worse than fetuses with microcystic lesions. Herein, we presented a fetus affected by macrocystic lung lesions with progressive hydropic changes during the second trimester, but experienced resolution of hydrops after a series of *in utero* interventions, eventually achieved livebirth and excellent outcome, in which the cystic lesions did not require surgical intervention.

Case report

A 19-year-old primigravida was referred to our tertiary medical center at 23^{+4} weeks of pregnancy because of cystic thoracic mass with hydropic change. At the first visit, the ultrasound revealed a huge multilocular cystic mass occupied the whole right lung and even caused the mediastinal-shift, left pleural effusion, and scalp edema. The lesions enclosed three cysts ranging from 16.4 mm to 18.6 mm in diameter (Fig. 1A and B), without apparent feeding vasculature from aorta. No additional structural anomalies were detected and the genetic investigations were offered, which showed 46, XX by amniocentesis.

Under the impression of macrocystic pulmonary lesions, the couple opted to continue pregnancy after non-directive genetic

☆ All the coauthors made significant contribution to the research and no conflict of interests exists.

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http://dx.doi.org/10.1016/j.tjog.2017.04.027

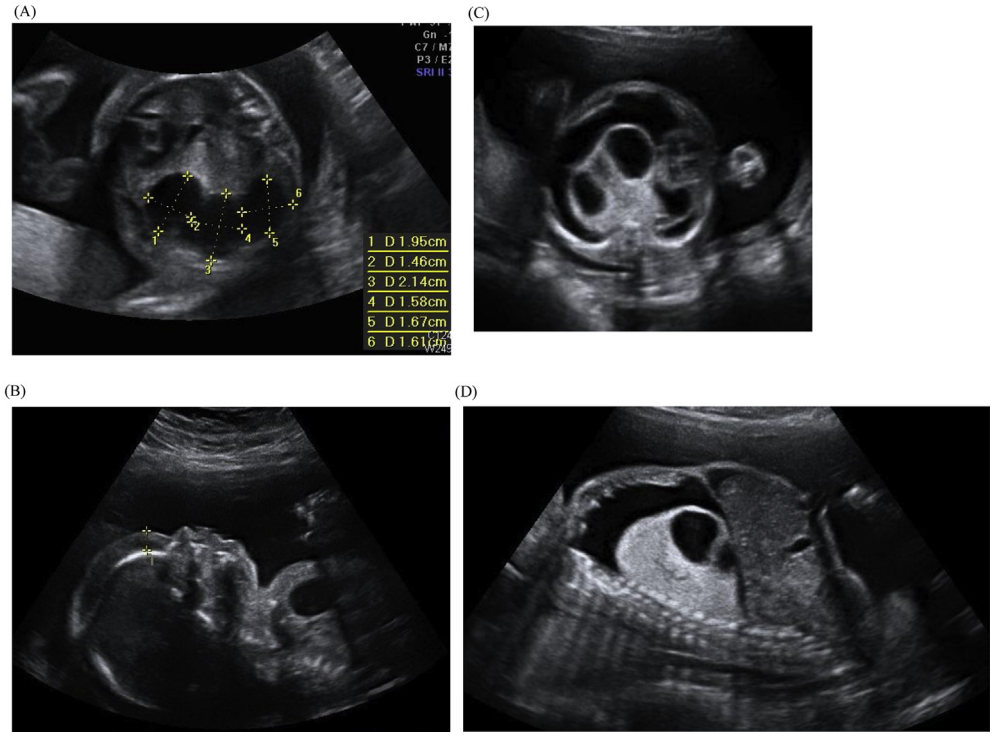

Fig. 1. (A) At 23^{+4} weeks of pregnancy, ultrasound showed a huge multilocular mass with three enclosed cystic lesions ranging from 16.4 mm to 18.6 mm in diameter occupied the right lung. (B) The fetus demonstrated scalp edema. (C) At 26 weeks of gestation, the fluids accumulated and lead to bilateral hydrothorax and ascites (D).

counseling. At 24 weeks of pregnancy, we performed intrauterine cyst aspiration followed by intra-cystic OK-432 injection at a dosage of 0.1 Klinische Einheit (KE; 1 KE = 1 mg/mL in the preparation from Chugai Pharmaceutical Co. Ltd., Tokyo, Japan). However, during the subsequent visits, the multilocular pulmonary cysts expended again and hydrops developed progressively, including bilateral pleural effusion, ascites and general skin edema (Fig. 1C and D). Poor prognosis was therefore informed according to our previous experience [3]. Thus again we offered rescuing thoracoamniotic shunting at 26th week of gestational age after pleurocentesis and paracentesis. Under ultrasound guidance, double basket shunt (shunt tube inner diameter 0.9 mm, outer diameter 1.47 mm; Hakko Co.) were inserted into the most accessible largest cyst followed by the second effort to place another catheter into the left lung for drainage of pleural effusion 10 days later. After that, we monitored the fetus by serial ultrasound examinations every week. Initially, the color Doppler of umbilical artery demonstrated absence of end diastolic velocity (AEDV), indicating arterial compensation due to the compression of the cysts onto the great vessels. Three weeks later, the shunts were in place and a remarkably resolution of the hydrops was noted (Fig. 2A and B), representing the excellent functioning of the catheters. Except for polyhydroamnios, the umbilical artery Doppler waveform became normal and the hydrops also resolved significantly. It indicated that the mass effect in the chest is the etiology of the hydrops and may be reversible. We also arranged prenatal magnetic resonance imaging (MRI) to exclude other anomalies to reassure the couple that the fetus should have a favorable outcome.

At 33^{+3} weeks of gestation, the baby was born via cesarean section due to preterm premature rupture of membranes and fetal malpresentation. A 1740-gm female infant without delayed of initial crying was born. The Apgar's scores was 5′ and 6′ at 1st min and 5th min (Fig. 2C). Initially, the newborn underwent endotracheal incubation and ventilator respiratory support. The postnatal image studies by contrast computer tomography presented an anomalous systemic artery arising from the descending aorta supplying the mass-like lesion with large cystic lesion in the right lower lobe of lung (Fig. 2D), indicating it is a case of pulmonary sequestration. The baby stayed in our neonatal units and then discharged 50 days later without any respiratory assistance. At five month old, she underwent thoracoscopic lobectomy, which revealed and confirmed that the lesion was intrathoracic extralobular lung sequestration.

Discussion

Congenital cystic lung malformations include a wide spectrum of pathological abnormalities: CCAMs, BPS, congenital lobar emphysema, and mediastinal teratoma [1]. The most common conditions are CCAMs and BPS [4], in which the major differential diagnostic feature is the origin of the feeding vasculature. CCAMs derive their blood supply from pulmonary vessels, as well as BPS is directly supplied from the descending aorta. However, owing to significant overlap in the findings and disease course of CCAMs and BPS, it is somewhat difficult for physicians to clearly delineate these two conditions at prenatal stage [5].

The management of congenital lung cystic lesions are mainly based upon if it is macrocystic or microcystic. Unlike fetuses with microcystic CCAMs tend to reach better outcomes since most lesions regress spontaneously after 26–28 weeks of gestation [5], macrocystic lesions of other etiologies generally do not regress and may carry a risk of pulmonary hypoplasia resulted from the compression effect of the large space-occupying lesions to the fetal lung parenchyma. Furthermore, when fetal hydrops occurs because of the compression effect to the inferior vena cava or even the heart itself by the accumulated pleural effusion and ascites. As a consequence, mortality rate increased dramatically. In one large retrospective study, all 25 hydropic fetuses died among 101 fetuses with CCAMs under expectant management [6], as well as another series involving 180 fetuses, only 3% of untreated hydropic fetuses survived,

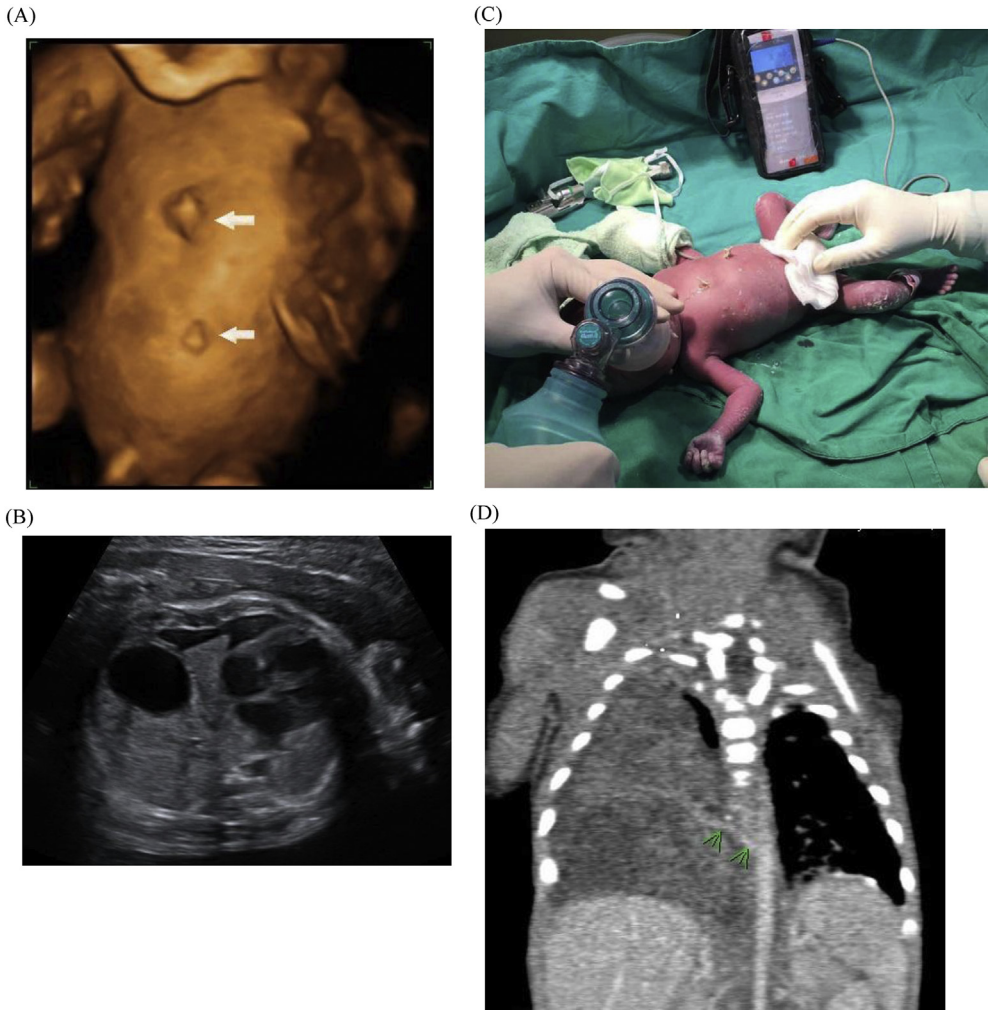

Fig. 2. (A) During the third trimester, the shunts were still in place demonstrated by 3D ultrasound (arrows). (B) Ultrasound performed at 33 weeks of gestation showing much-alleviated hydrops and minimal left pleural effusion. (C) At birth, the shunts were in place. (D) Postnatal chest computed tomography demonstrated a feeding vessel (arrows) originating from the descending aorta and supplying the mass-like lesion, indicating it is a pulmonary sequestration.

compared to 97.6% of non-hydropic fetuses [7]. Thus, prenatal interventions, including steroid therapy, thoracocentesis, thoracoamniotic shunting, laser or radiofrequency ablation of the feeding vessels, and open fetal surgery are reported to treat hydropic fetuses due to lung lesions and massive pleural effusion [8].

Thoracoamniotic shunting is a feasible option because the risk of fetal demise associated with hydrops outweighs the risk of the procedure itself if the shunting procedure being performed in an expert's hand. A recent systematic review showed an improved survival rate of 62% in treated hydropic fetuses vs. 3% in those untreated (OR, 19.28; 95% CI, 3.7–101) [9]. Whereas, limited studies has reported solely on macrocytic lung lesions. To date, the largest one reported the outcome of thoracoamniotic shunting on 68 fetuses with macrocystic CCAMs was by Cavoretto and the colleagues [10], followed by other two small series, each enrolled 11 cases [11,12]. The overall survival rate was 75%, 68% and 83% in hydrops, respectively and 87.5% in non-hydropic fetuses.

In our case presented here, it demonstrated several poor prognostic markers before 28 weeks of gestation, including huge size, macrocytic lung lesions, the presence of mediastinal shift, polyhydramnios, and especially hydrops (which is highly associated with stillbirth) [12]. Hence, the aim of prenatal intervention is to alleviate the mass effect generated by the cystic lesions to the fetal lungs, the great vessels, and eventually the fetal heart, to continue the pregnancy into the third trimester, when the mass size tended to minimize and better neonatal outcomes could possibly be achieved. Despite the hydrops didn't resolve immediately after shunt placement. Eventually, the fetus recovered three weeks later, and reached a remarkably favorable outcome, which echoes well the prior reports that thoracoamniotic shunting should be offered for all hydropic fetuses with large macrocystic lung lesions [12] by skilled experts. In conclusion, we described a successful prenatal intervention by minimally invasive method to treat the macrocystic lung lesion coexisting with hydrops and argued that thoracoamniotic shunting is a preferred option for similar cases to enhance perinatal survival, especially adopting the delicate Japanese-designed double-basket catheter [13].

Acknowledgment

This study was partly funded by the grant from Ministry of Science and Technology, Executive Yuan, Taiwan, R.O.C. (MOST 101-2314-B-371-004-MY3) to M Chen.

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