CASE REPORT

Refractory tension pneumothorax as a result of an internally displaced thoracoamniotic shunt in an infant with a congenital pulmonary airway malformation

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Accepted 13 July 2016

SUMMARY

Antenatally, congenital pulmonary airway malformation (CPAM) causing fetal hydrops can be palliated with thoracoamniotic shunts, which may become displaced in utero. We report a case of an infant born at 34 weeks gestational age with an antenatally diagnosed macrocystic lung lesion, fetal hydrops and an internally displaced thoracoamniotic shunt. The infant suffered refractory pneumothoraces despite multiple chest drains, and stabilised only after surgical resection of the lesion. Intraoperatively, the shunt was noted to form a connection between a type I CPAM and the pleural space. As the shunt was displaced internally, this complication was not immediately obvious during the initial resuscitation. In infants with large cystic lung lesions, clinicians should be aware that internally displaced thoracoamniotic shunts could contribute to refractory tension pneumothoraces and anticipate the need for advanced neonatal resuscitation, including early thoracocentesis or chest drain insertion. Furthermore, displaced shunts may require early surgical intervention.

BACKGROUND

As antenatal ultrasound scans have become a part of standard prenatal care, cystic lung lesions are sometimes identified, and can represent a range of congenital thoracic malformations (CTMs), including congenital pulmonary airway malformations (CPAMs).^{1 2} While many of these malformations are well tolerated in utero, cystic lesions that are significant in size may cause fetal hydrops, necessitating antenatal interventions such as the ultrasound-guided thoracocentesis and the placement of thoracoamniotic shunts. In particular thoracoamniotic shunts can improve fetal hydrops, but may also complicate neonatal management of these infants.^{3–6}

CASE PRESENTATION

A baby girl was born to a 30-year-old primigravida mother. The pregnancy was initially without complications or exposures. A left macrocystic lung lesion was first seen on an antenatal ultrasound scan at 19+ weeks gestation, measuring 2.1×1.0×1.3 cm. Otherwise fetal anatomy appeared normal. At 24+ weeks, an ultrasound scan demonstrated a progressively enlarging cyst (6.7×6.6×3.5 cm) with associated signs of fetal hydrops, including mild ascites and left-sided pleural effusion. Owing to early signs of hydrops,

the cyst was aspirated at this time under ultrasound guidance. Fetal echocardiogram demonstrated normal biventricular function, and left pulmonary artery diameter at the lower limit of normal. Two subsequent aspirations were performed, as the cyst reaccumulated. At 26+ weeks, an ultrasound scan again demonstrated mild-to-moderate ascites, pleural effusion and continued expansion of the cyst, reaching a size of 7.9×6.2×4.2 cm (figure 1A). As a result, a thoracoamniotic shunt was inserted under ultrasound guidance, draining from the cyst into the amniotic fluid, resulting in successful decompression of the cyst and resolution of hydrops (figure 1B, C).

However, at 28 weeks of gestation, the cyst had reaccumulated to 5.8×4.9×3.3 cm. The shunt appeared to have been displaced into the amniotic cavity. A second shunt was placed, which continued to decompress the cyst until 31 weeks gestation. Subsequently, fluid re-accumulated within the lesion, and the shunt appeared to have been displaced internally into the cyst. Serial ultrasound scans performed between 31 and 33 weeks demonstrated steady enlargement of the cyst, reaching a size of 5.1×2.7×3.9 cm. Mild left pleural effusion without ascites was attributed to either early hydrops or fluid leakage from the cysts into the pleural cavity. As a result of this, and of increasing maternal discomfort due to maternal habitus, a caesarean section was performed at 34 weeks of gestation after administration of steroids.

Prior to delivery, fetal heart rate was reassuring. A primary lower transverse segment caesarean section was performed under general anaesthesia for maternal indications. The neonatal resuscitation team was present at the time of birth. Given the known history of a large macrocystic lung lesion and history of displaced thoracoamniotic shunts, prior to the delivery preparations were made for emergency needle thoracocentesis after birth in anticipation of a possible pneumothorax. The infant was unexpectedly apnoeic and hypotonic and was shifted immediately to the resuscitation room. No heart rate could be palpated or auscultated despite adequate bag–mask ventilation; she was intubated at 2 min and received 1-min of chest compressions. With intubation and ventilation, the heart rate rapidly improved and thoracocentesis was not performed. She remained haemodynamically stable but hypotonic, with pinpoint pupils and lacking gag

To cite: Law BHY, Bratu I, Jain V, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-216324

Figure 1 Antenatal ultrasound scans. (A): large left chest cystic lesion (*) is seen displacing the heart (white arrow) in the four-chamber view, with minimal lung tissue visible on either side. (B): the shunt (black arrow) is seen in the cyst during ultrasound-guided placement. (C): normalisation of the four-chamber view of the heart (white arrow) the day after shunt placement, a small secondary cyst is seen near the cardiac apex after complete decompression of the primary.

response or response to physical stimuli. APGARs were 0, 3 and 4 at 1, 5 and 10 min respectively. The birth weight was 2380 g. Cord gases were normal, with an arterial pH of 7.3 and a base deficit of 2.1. On inspection of the chest wall, no shunt or defect was seen. She remained easy to ventilate on conventional ventilation and was transferred to the neonatal intensive care unit (NICU) for further stabilisation.

Umbilical arterial and venous lines were placed in preparation for transport to the surgical NICU. The first obtained chest radiograph after central line insertion revealed a left pneumothorax, a rightward mediastinal shift and a large ($\sim 5 \times 4$ cm) cystic lesion within the left lung. A thoracoamniotic shunt was clearly visible within the cyst (figure 2). While initially stable postresuscitation, mechanical ventilation became increasingly difficult during the first 2 hours of life; target tidal volumes could not be achieved despite high inspiratory pressures, and oxygen needs increased to 0.5 Fractional Inspired Oxygen (FiO_2). She did not develop hypotension despite the pneumothorax and mediastinal shift. At 2 hours of life, given the worsening respiratory status and a radiograph confirming a large pneumothorax, a pigtail catheter was inserted in the left chest to relieve the pneumothorax, initially draining 35 mL of air and 5 mL of fluid. Her neurological and ventilation status improved and she was then transported to the surgical NICU for further management.

While the infant was <36 weeks of gestation age, in consultation with a second neonatologist, therapeutic cooling was initiated for neuroprotection given an abnormal neurological

status consistent with stage II hypoxic-ischaemic encephalopathy and the need for cardiac compressions at birth. She subsequently developed recurrent tension pneumothorax, with associated hypotension and ventilation difficulties despite the placement of additional chest drains, fluid resuscitation and inotropes. She was brought to the operating room on the second day of life for a thoracotomy and left upper lobectomy and removal of the lesion. Pathology confirmed a type I CPAM located in the left upper lobe measuring $5.1 \times 4 \times 1$ cm. Intraoperatively, the second thoracoamniotic shunt was found to be only partially displaced into the cyst, forming a connection between the cyst and the pleural cavity.

OUTCOME AND FOLLOW-UP

Postoperatively, the infant recovered remarkably well, and was successfully extubated on postoperative day 4. Her neurological status improved, and her brain MRI was normal. She was transferred on the eighth day of life to a community NICU for further management of her prematurity and discharged home on the 16th day of life on oral feeds and no respiratory support. She remained well on follow-up at 9 weeks. At 6 months, her mother described her as strong and healthy and gave consent to publish this case report.

DISCUSSION

CTM are a heterogeneous group of rare malformations, which include CPAMs, bronchogenic cysts, pulmonary sequestrations and congenital lobar emphysema and other cystic lesions.^{1 2}

Figure 2 Initial postnatal chest radiograph showing left-sided tension pneumothorax, cystic lung lesion and an internally retained thoracoamniotic shunt.

With improvement in ultrasound imaging technology, these lesions are often visualised antenatally.⁶ While various classification schemes have been employed to describe these lesions, they are usually difficult to differentiate based on an ultrasound scan alone, and are usually described based on ultrasonographical appearance.⁵

Antenatally, cystic lung lesions can rarely cause significant intrathoracic mass effect, resulting in pulmonary hypoplasia, mediastinal shift and hydrops fetalis. Hydrops fetalis and increased congenital pulmonary malformation volume ratio have been shown to be associated with neonatal morbidity and mortality; prior to the era of fetal interventions, the natural history of cystic lung lesions with hydrops fetalis was almost universal death or fetal demise.^{4–7} Systematic reviews have demonstrated that antenatal cyst decompression convey a survival benefit in those cases with fetal hydrops; however, the available studies tend to be of small numbers with variable techniques, lung lesions and follow-up, and none are randomised control trials.^{7–8} Fetal thoracoamniotic shunts have been successfully placed to decompress these lesions and may reverse existing hydrops.^{7–11} While institutional experiences with fetal thoracoamniotic shunts differ, previous case series have demonstrated postnatal survival of close to 70%.^{9–11} Known perinatal complications of shunt insertion include membrane separation, preterm labour and bleeding, as well as shunt malfunctions such as dislodgement or occlusion. Postnatally, there has also been an association with shunt placement and chest wall abnormalities.¹²

While most infants with antenatally diagnosed CTMs are well, for infants with large lesions, the postnatal course can be complicated by lung hypoplasia and pneumothorax. Infants with CPAM and hydrops fetalis are at particular risk; the postnatal mortality

of live-born infants with CPAM complicated by hydrops has been reported to be 31%.⁷ Delivery at a tertiary care centre is indicated and clinicians should anticipate the need for advanced neonatal resuscitation, including early thoracocentesis or chest drain placement. The presence of thoracoamniotic shunts can further complicate postnatal management. A shunt visible at the chest wall can clearly be the cause of a pneumothorax and should be promptly removed or clamped and a chest drain promptly inserted; an internally displaced shunt is a less obvious culprit. In our case, refractory pneumothorax was only resolved when the cystic lesion was excised and the shunt was found to form a communication between the cyst and the pleural space, acting as an artificial bronchopleural fistula. As the infant's lungs were ventilated, air was directed rapidly into the pleural space, resulting in refractory tension pneumothorax. The frequency and severity of complications related to internally displaced thoracoamniotic shunts is not well known; cases reported in the literature describe a wide range of outcomes from asymptomatic to severe complications, mostly related to displaced pleuroamniotic shunts in the treatment of fetal hydrothorax.^{13–16} In one case, there was neonatal demise related to the displacement of pleural-amniotic shunt within the pleural cavity causing hilar compression.¹⁴ To the best of our knowledge, a cyst–pleural communication formed by a displaced thoracoamniotic shunt resulting in refractory tension pneumothorax has not yet been described. Clinicians involved in neonatal resuscitation and management of infants with antenatally placed thoracoamniotic shunts should be aware of the risk of pneumothorax, even in the absence of externally visible shunts or chest wall defects. In case of internally displaced thoracoamniotic shunts, immediate needle thoracocentesis or chest drain insertion should be strongly considered after birth to relieve any tension pneumothorax. If a thoracoamniotic shunt is suspected to be internally displaced on antenatal ultrasound scan, clinicians attending the birth of an infant should be made aware of the displaced shunt and advised to consider immediate thoracocentesis or chest drain insertion postbirth. The insertion of postnatal chest drains may be insufficient; earlier surgical intervention may be necessary. Thus, paediatric surgery should be involved early in the management of an infant with an internally displaced thoracoamniotic shunt.

Learning points

- ▶ In neonates with antenatally diagnosed macrocystic lung lesions, antenatally placed thoracoamniotic shunts may become internally displaced. While they may not be seen at birth, displaced shunts may form a communication between the lesion and the pleural cavity, contributing to refractory pneumothorax.
- ▶ Clinicians should anticipate the need for an advanced resuscitation and strongly consider early drainage of pneumothoraces when a history internal shunt displacement has been documented.
- ▶ In infants with large congenital pulmonary airway malformations, internally displaced thoracoamniotic shunts may necessitate early surgical intervention.

Contributors BHYL conceptualised this report, gathered the data, drafted the manuscript and approved the manuscript as submitted. M-AL aided in the conceptualisation of this report, critically reviewed and approved the manuscript as submitted. IB and VJ provided additional data, critically reviewed, revised and approved the manuscript as submitted.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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