Pneumothorax associated with a displaced thoracoamniotic Somatex shunt in an infant with congenital pulmonary airway malformation: a case report

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Case presentation

A 32-year-old nulliparous pregnant woman at 21 weeks of gestation was referred to Kwong Wah Hospital in March 2021 for fetal right cystic lung mass (2.16×1.99×2.50 cm^3). Repeat examination at 22 weeks of gestation revealed a right multicystic lung mass (3.46×2.46×2.37 cm^3) with a dominant 2-cm cyst, suggestive of macrocystic congenital pulmonary airway malformation (CPAM). There was mild mediastinal shift but no hydrops. The CPAM volume ratio, calculated as (length×height×width×0.52)/head circumference, was 0.51. Amniocentesis with chromosomal microarray analysis showed no copy number variants. At 26 weeks of gestation, the lesion had enlarged to 5.73×4.28×4.26 cm^3 (CVR=2.16), with mediastinal shift and mild ascites but no polyhydramnios. At 27 weeks of gestation, the lesion was dominated by a single cyst that measured 6.15×4.25×4.1 cm^3 (CPAM volume ratio=2.09), with moderate polyhydramnios, ascites and skin oedema suggestive of fetal hydrops. Intramuscular betamethasone was administered for fetal lung maturation in view of the high risk of preterm delivery. Fetal thoracoamniotic shunting was offered to relieve the mass effect and fetal hydrops. The next day, a Somatex shunt (SOMATEX Medical Technologies, Berlin, Germany) was inserted into the CPAM under ultrasound guidance and 800 mL amniotic fluid was drained via the shunt cannula. Examination 9 days later showed CPAM with reduced size (2.3×1.7×1.5 cm^3; CVR=0.12), shunt in situ, and no hydrops (Table).

At 29 weeks of gestation, the patient went into preterm prelabour with rupture of membrane that sealed off spontaneously. Serial ultrasound examinations showed satisfactory fetal growth, collapsed CPAM with shunt in situ, normal liquor volume and no hydrops (Table). Labour was induced at 38 weeks of gestation for oligohydramnios. A 2.75-kg female baby was delivered by vacuum extraction for maternal exhaustion, with paediatrician standby. The Apgar scores of the baby were 8 at 1 minute and 10 at 5 minutes and the arterial cord blood pH was 7.31, with base excess of -7.1 mmol/L. The shunt was specifically searched for immediately after delivery but the skin was intact (Fig a). The baby was given continuous positive airway pressure because of respiratory distress and was transferred to the neonatal intensive care unit. Urgent chest X-ray revealed the shunt in the right chest with right pneumothorax (Fig b). A chest drain was inserted and the baby was intubated. Computed tomography of the thorax of the baby on day 1 of life showed an irregular 4.2×3.5×2.5 cm^3 cystic lesion in the right lung with the distal end of the shunt migrated between the chest wall and the scapular, abutting the right subscapularis muscle. Thoracoscopy on day 6 of life confirmed that one end of the shunt was within the CPAM in the right middle lobe, while the other end was at the subscapular space. The shunt was removed intact and near-total right middle lobe excision was performed thoracoscopically. The baby was successfully weaned off oxygen 3 weeks postoperatively and discharged 5 weeks later.

Discussion

Congenital pulmonary airway malformations are uncommon lung lesions characterised by an overgrowth of terminal respiratory bronchioles that are often immature and non-functioning. A CPAM is considered macrocystic if at least one cyst is >5 mm and microcystic if the lesion appears echogenic on ultrasound examination. Although a microcystic CPAM may regress spontaneously after 26 to 28

TABLE. Antenatal course of the congenital pulmonary airway malformation of the baby

Gestation, wk	CPAM size	CVR	Hydropic change	Liquor volume
22^{+2}	3.46×2.46×2.37 cm^3 (with 2-cm dominant cyst)	0.51	No	Normal
23^{+5}	3.09×2.63×3.6 cm^3	0.67	No	Normal
26^{+2}	5.73×4.28×4.26 cm^3	2.16	Mediastinal shift, thin rim of ascites, no skin oedema	Normal
27^{+4}	6.15×4.25×4.1 cm^3 (single macrocyst)	2.09	Thin rim of ascites, skin oedema, mediastinal shift	Polyhydramnios (AFI=33.7 cm)
29^{+1}	2.3×1.7×1.5 cm^3; shunt* in situ	0.12	Resolved, mild mediastinal shift	Normal (AFI=24 cm)
31^{+2}	Variable, from 1.5 cm to 2.5 cm in diameter; shunt in situ with bidirectional flow	N/A	Resolved	Normal
32^{+1}	Almost completely collapsed; shunt in situ	N/A	Resolved	Normal (AFI=9.7 cm)
33^{+2}	Collapsed; shunt in situ	N/A	Resolved	Slightly reduced (AFI=7.2 cm)
35^{+2}	Collapsed; shunt in situ	N/A	Resolved	Normal
36^{+2}	Collapsed; shunt in situ	N/A	Resolved	Normal
37^{+2}	Collapsed; shunt in situ	N/A	Resolved	Normal

Abbreviations: AFI = amniotic fluid index; CPAM= congenital pulmonary airway malformation; CVR = congenital pulmonary airway malformation volume ratio; N/A = not applicable

* Somatex thoracoamniotic shunt was inserted into the CPAM under ultrasound guidance at 27^{+6} weeks of gestation

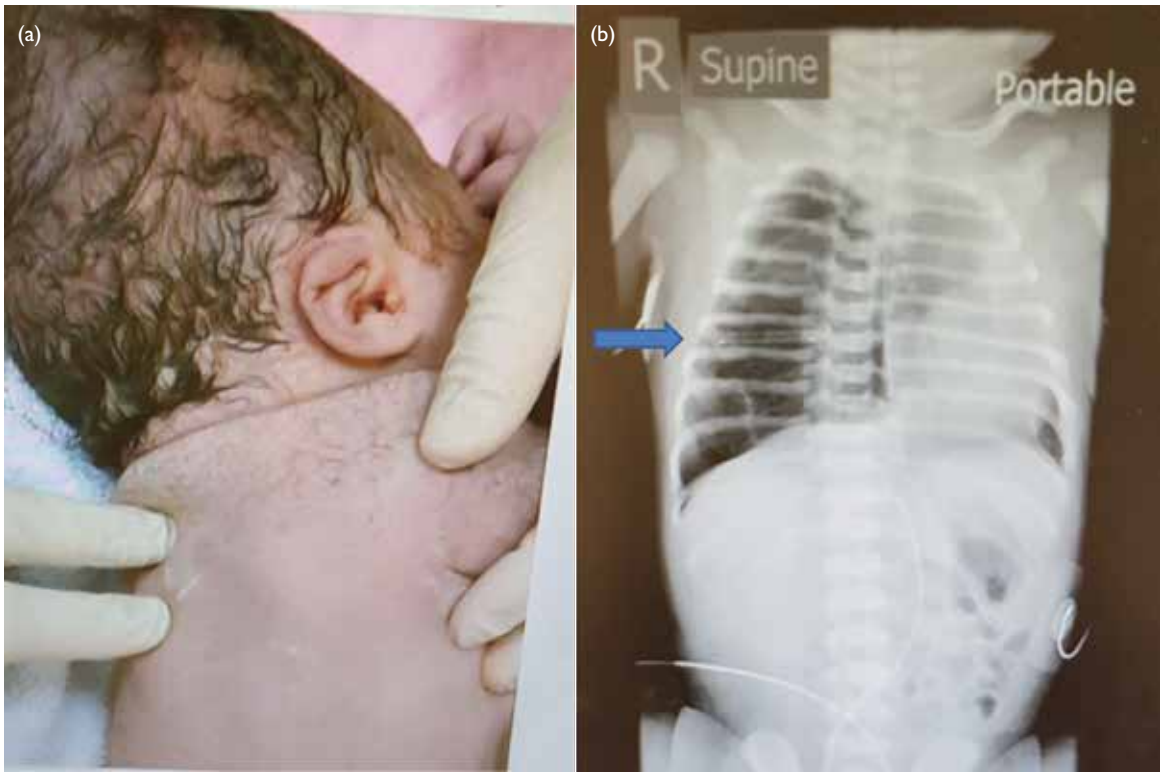

FIG. (a) Intact skin overlying the right chest wall of the newborn immediately after birth. (b) Chest X-ray showing the internally displaced Somatex shunt in the right middle lobe (arrow), with pneumothorax

weeks of gestation, most macrocystic CPAMs do not.[1] Fetuses with large cystic lesions are also at risk of pulmonary hypoplasia and development of fetal hydrops due to compression of lung tissue and venous return. The survival of a hydropic fetus with congenital lung lesion has been reported to be 38%, compared with 87% for a fetus without hydrops.[1] Studies have demonstrated a favourable outcome following thoracoamniotic shunting for macrocystic CPAM, with reduction in lesion volume, resolution of hydrops and improved survival.[2,3] A systematic review showed an improved survival from 3% to 62% in hydropic fetuses treated with shunting.[4] In a single-centre case series, survival was significantly associated with gestational age at birth, hydrops resolution and higher percent reduction in the size of the lung lesion following shunting.[3]

Although thoracoamniotic shunting improves fetal outcome, complications such as shunt occlusion, displacement, dislodgement, bleeding and chest wall deformation have been reported.[3,5-7] Following successful drainage of the lesion, its surrounding normal lung parenchyma expands and grows. This may result in inward migration of the shunt. In a retrospective review,[5] thoracoamniotic shunts inserted for primary pleural effusions and macrocystic CPAMs were antenatally displaced in 8.5% of fetuses, of which two-thirds migrated into the thorax. Re-shunting may be required if the displaced shunt fails to drain and fluid re-accumulates.[5] Retained intrathoracic shunts may be managed conservatively as they are well tolerated without untoward postnatal sequalae.[5,8] Nonetheless surgical removal may be necessary if the baby develops complications such as respiratory distress or tension pneumothorax.[7,9]

The Somatex shunt is commonly used to treat fetuses with obstructive urinary tract disorders. Recently, thoracoamniotic shunting with a Somatex shunt has been reported effective in relieving fetal pleural effusions with good survival rate although shunt dislodgement and entrapment has been reported in four of eight cases.[10] Thoracoscopic removal of a displaced Somatex shunt has been reported necessary in a newborn with respiratory distress and progressive pleural effusion.[7] In comparison with other commonly used shunts, such as Harrison and Rocket, Somatex insertion has multiple advantages including a finer introducer (1.2 mm) but a bigger shunt lumen (2.4 mm). It is also made of metal facilitating its easy identification antenatally on ultrasound or postnatally with X-rays or computed tomography.

A thoracoamniotic shunt should be clamped immediately following delivery to prevent air from entering the thorax and causing pneumothorax.[2] In the current case, we did not expect air to enter the pleural cavity because the shunt was buried inside the skin of the baby. Our hypothesis is that air may have entered from the lung tissue into the pleural space via the displaced shunt. This is similar to the reported case of tension pneumothorax due to an internally displaced thoracoamniotic shunt communicating between the CPAM and the pleural cavity and diagnosed following neonatal resuscitation for apnoea.[9] Both cases illustrate that pneumothorax is a possible and potentially life-threatening complication of an internally displaced shunt. It should be anticipated at birth and preparations made for emergency needle thoracocentesis. Obstetricians should be aware of the possible complications of thoracoamniotic shunts, and paediatricians should be alerted so that the newborn can receive prompt assessment and treatment.

Author contributions

Concept or design: VYT Chan, WC Leung, TY Leung.
Acquisition of data: VYT Chan, WT Tse, MC Chan, KKY Wong.
Analysis or interpretation of data: VYT Chan.
Drafting of the manuscript: All authors.
Critical revision of the manuscript for important intellectual content: KKY Wong, WC Leung, TY Leung.

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

As an editor of the journal, KKY Wong was not involved in the peer review process. Other authors have disclosed no conflicts of interest.

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Ethics approval

The patient and her baby were treated in accordance with the Declaration of Helsinki. Parental consent was obtained for the patient's baby, and informed consent was obtained from the patient for all treatments and procedures, and publication of the case report.

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