Received: 23 January 2024 | Revised: 26 April 2024 | Accepted: 3 May 2024

DOI: 10.1002/ccr3.8956

CASE REPORT

Clinical Case Reports Open Access WILEY

Neonatal pleural effusion associated with pulmonary sequestration: A case report

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Key Clinical Message

Pleural effusion is rare during neonatal period with an estimated prevalence of 0.06%. It may sometimes uncommonly be secondary to pulmonary sequestration. Besides common conditions like hydrops fetalis, congenital heart disease, congenital chylothorax, chromosomal abnormalities; pulmonary sequestration should also be considered while evaluating the cause for neonatal pleural effusion.

KEYWORDS

extralobar, neonate, pleural effusion, pulmonary sequestration, respiratory distress

1 | INTRODUCTION

Pleural effusion is an abnormal collection of fluid within pleural space usually resulting from excess fluid production and or decreased lymphatic absorption. Pleural effusion is rare during neonatal period with the estimated prevalence of 0.06%.[1] Neonatal pleural effusion may be due to antenatal congenital causes or postnatal acquired causes. Antenatal pleural effusion may be due to hydrops fetalis, congenital chylothorax, congenital heart disease (CHD), chromosomal anomalies, pulmonary anomalies, or infections like congenital herpes simplex viral (HSV) and Parvo virus infection. Structural fetal malformations like congenital pulmonary airway malformations (CPAM), bronchopulmonary sequestration, congenital diaphgramatic hernia can also cause neonatal pleural effusion. Postnatal acquired causes include extravasation of peripherally inserted central catheters, parapneumonic effusion or traumatic chylothorax.

Pulmonary sequestration is very uncommon abnormality of lower respiratory tract. It is classified as intralobar and extralobar types based on the relationship with the lung parenchyma. Pulmonary effusion secondary to extra lobar pulmonary sequestration is a very rare entity.[2]

As neonatal pleural effusion can cause significant respiratory distress, vigilant delivery room management is required. Identifying the underlying cause is important as it leads to specific therapeutic measures. Diagnostic chests tap for pleural fluid analysis is important for diagnosis and its consequent management.[3] Similarly, echocardiography, karyotyping and other investigations are also indicated for identifying subsequent etiology.

Here, we report a case delivered at 36 + 2 weeks of gestation (WOG) with antenatal scan showing polyhydramnios and left sided pleural effusion, which was later diagnosed as extra lobar pulmonary sequestration after evaluation for all possible explanations. The rarity of this clinical condition make this case report a valuable asset to the scientific literature. The purpose of this case report is to familiarize clinician regarding lung sequestration as the cause of neonatal pleural effusion and prevent subsequent morbidity and mortality.

2 | CASE REPORT

A 22 years old G2P1L1 healthy female from Dang district of Nepal presented at 36 + 2 WOG with leaking and labor pain. The antenatal scan was only done at 32 WOG which showed polyhydramnios in mother with fetal left sided pleural effusion. She was referred to Patan hospital for further evaluation and management. At Patan hospital ultrasonography (USG) scan was repeated which reconfirmed left sided pleural effusion without any other associated congenital anomalies. She delivered male baby weighing 2290 g which is appropriate for gestational age (AGA) via spontaneous vaginal delivery with an apgar score of 7/10 and 8/10 at 1 and 5 min, respectively. Baby developed grunting, sub costal retractions at 30 min of life. Baby was transferred to Neonatal Intensive Care Unit (NICU).

At NICU, baby was tachypenic with respiratory rate of 65 per min and mild sub costal retractions with SPO_2 of 80% at room air; Downe's score was 4/10. There was no dysmorphism or any other congenital anomalies. Air entry was decreased on left lung. Cardiovascular examination was normal. There was no sign of heart failure. Abdomen was soft and there was no sign of fluid in the abdominal cavity. Childreceived mechanical ventilation in nasal-synchronized intermittent mandatory ventilation (SIMV) mode. Emperic antibiotic was started after obtaining culture. Chest x-ray was done (Figure 1) which showed left sided pleural effusion. Chest tube was inserted (Figure 2) and approximately 200 mL of pleural fluid was aspirated. Aspirated sample was sent for analysis.

3 | METHODS

Hydrops fetalis, congenital chylothorax, congestive cardiac failure, fetal aneuploidy, structural fetal malformations like CPAM, bronchopulmonary sequestration, congenital diaphgramatic hernia were sought as the possible differential diagnosis.

Evaluation to establish the cause began with pleural fluid analysis. Findings with the normal reference is shown in Table 1. Pleural fluid was straw colored. Pleural fluid biochemistry showed protein 1.2 g/dL sugar 97 g/dL lactate dehydrogenase (LDH) 82 U/L triglyceride 13 g/dL total cholesterol 50 g/dL. Microscopic examination showed red blood cell (RBC) count: 10/mm^3, white blood cell (WBC) count 100/mm^3 with polymorph of 30% and lymphocyte of 70%.

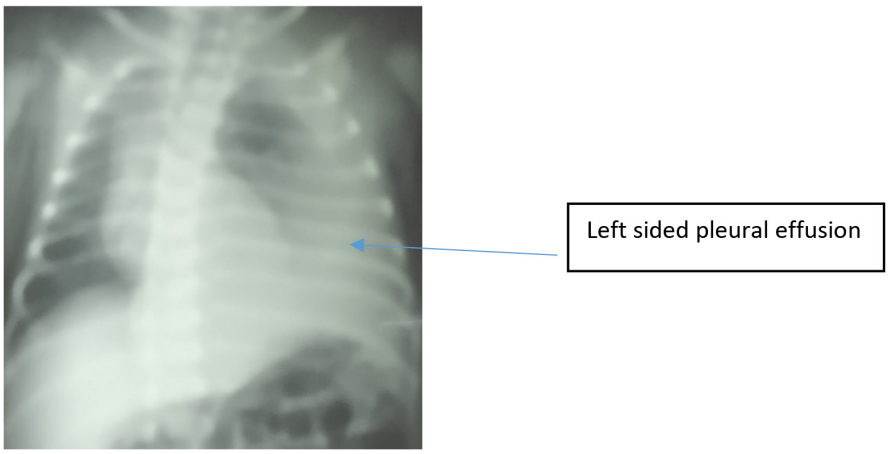

FIGURE 1 Chest x-ray AP view after delivery showing left sided effusion.

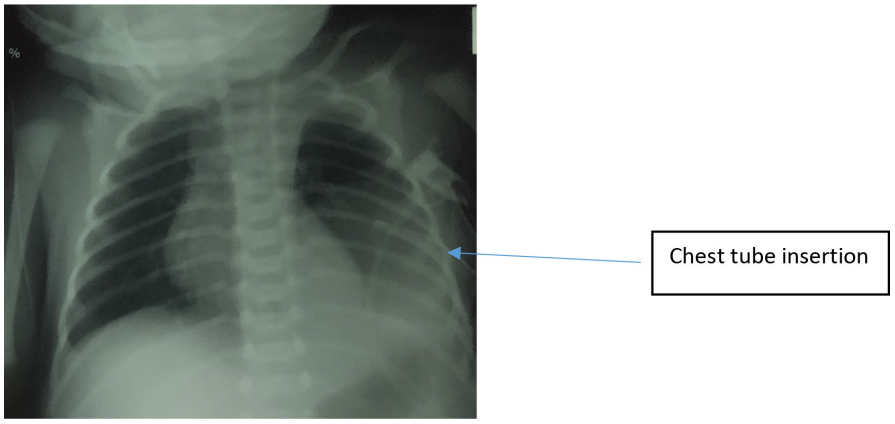

FIGURE 2 Chest x-ray AP view after chest tube insertion.

As the pleural fluid triglyceride level is less than 110 mg/dL and the ratio of pleural fluid protein and serum protein less than 0.5, ratio of pleural fluid LDH and serum LDH less than 0.6; pleural fluid was found to be non-chylous and transudative in nature.

Pleural and blood culture were sterile. Echocardiography was done which showed small atrial septal defect (ASD); left to right shunt. No pericardial effusion and no signs of cardiac failure. Urine examination was normal with no proteinuria. Ultrasonography revealed minimal effusion in the lung and possibility of pulmonary sequestration. Ascites was absent. Contrast enhanced computed tomography (CECT) chest was done (Figure 3) and final diagnosis of extralobar pulmonary sequestration (ELS) was made. As the cause of pleural effusion was identified, karyotyping wasn't done.

Child's condition improved following chest tube drainage. Respiratory rate was within normal limits (52 breaths per min), no sign of respiratory distress (no grunting, no chest retractions), with saturation 92%. Capillary blood gas was within normal limits; pH 7.36 (7.35–7.45), PCO_2 mmHg (35–45 mmHg) bicarbonate 25 mmol/L (22–28 mmol/L). Respiratory support gradually weaned to room air. Repeat chest x-ray did not show fluid reaccumulation. Hence, chest tube was removed. Antibiotics was stopped once blood culture was sterile. Child was transferred to nursery after 7 days. Cardiothoracic and vascular surgery (CTVS) consultation was done. The child was advised for follow up at 3 months for further evaluation and planning for next course of action.

TABLE 1 Pleural fluid analysis.

Characteristics	Results	Normal reference
Color	Straw colored	Colorless
WBC count	100/mm^3	<1000/mm^3
Lymphocytes	70%	5%
Triglycerides	13 g/dL	<50 mg/dL
Protein	1.2 g/dL	1–2 g/dL

Note: Blood workup was done; findings are enlisted in Table 2.

4 | DISCUSSION

Pulmonary sequestration is a congenital lesion that consists of anomalous lung parenchyma that has own arterial supply with no connection to tracheobronchial tree. This rare abnormality has an incidence between 0.15% and 6.45% among all pulmonary malformations.[4] It is divided two intralobar (ILS) and ELS. Intralobar sequestration is contained within normal lung parenchyma with venous drainage to pulmonary vein whereas extralobar is separated from normal lung and has its own visceral pleura and has venous drainage to systemic vein.[5]

ELS accounts for only 25%–15% of all sequestration.[6] Most ELS present in the first 6 months of life with one-quarter of babies presenting shortly after birth with respiratory distress or feeding difficulties.[7] Slightly older children present with respiratory symptoms and occasionally with congestive cardiac failure. It can be seen on fetal ultrasound as early as 18 WOG as a well-defined triangular echogenic mass in the lower chest or the suprarenal region of the abdomen.[8] However, some may of them involute spontaneously and even disappear. Antenatal diagnosis provide an opportunity for perinatologist to decide whether to interrupt pregnancy, treat complication in utero or initiate delivery of a fetus in distress. Establishment of systemic arterial supply and venous drainage of sequestrated lung tissue is necessary for definitive diagnosis. Multi-planar CT with three dimensional reconstruction is currently preferred non-invasive investigation of choice currently compared to digital subtraction angiography. Magnetic resonance angiography is also a reasonable alternative.[9]

TABLE 2 Laboratory parameter.

Parameters	Value	Reference
Total leucocyte count	15,000/mm^3	9.1–34.0 × 1000/mm^3
Neutrophil	35%	54%–62%
LDH	750 U/L	170–580 U/L
Protein	5 g/dL	4.6–7.4 g/dL

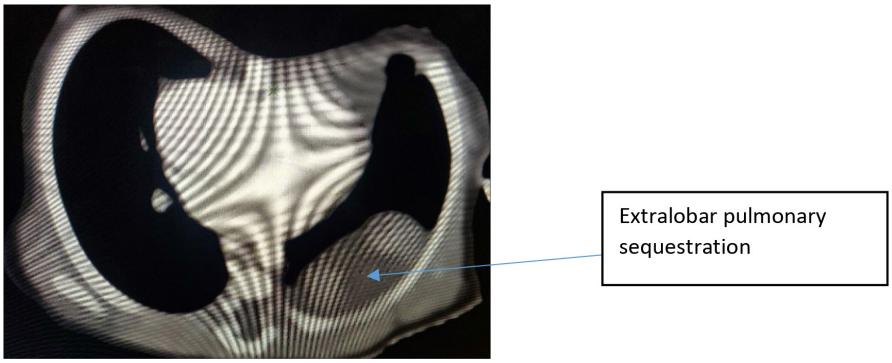

FIGURE 3 Contrast enhanced computed tomography (CECT) chest showing extralobar pulmonary sequestration (ELS).

Neonatal pleural effusion secondary to ELS has rarely been reported in the literature. Earlier, Bliek AJ et al.,[10] Dresler S,[11] Horowitz RN,[12] J Lukaya et al.[2] have reported this unusual clinical condition. Following were the findings of those case reported.

Table 3 shows comparison of all the previous similar case reports. Neonates in those case reports were born premature at or before 34 WOG. All of them developed respiratory distress immediately at birth and required vigorous resuscitation at delivery room. However, none could be saved and diagnosis was made based on the post mortem findings. In contrast, the neonate in our case was delivered late preterm at 36 + 2 WOG. Baby developed respiratory distress but did not require vigorous resuscitation. Similarly, thoracostomy to drain pleural fluid was done immediately which might have led to a favorable prognosis in contrary to other cases. Diagnosis on our neonate was done based on USG and CECT scan findings. Though ultrasound plays an important role in prenatal diagnosis and follow-up of pulmonary sequestration as aberrant vessel of feeding vessel of lung mass can be identified; findings are non-specific and has limited value.[13] Extralobar sequestration on CT appears as a well-defined mass of uniform soft-tissue attenuation often with single anomalous artery arising from the thoracic or abdominal aorta and it almost never contains air.[14] Baby was discharged after improvement. Anomaly in our case was left sided, which was consistent with all other reported neonates. Polyhydraminos has been reported in our case along with two of the other studies. Polyhydraminos has been associated with the poor prognosis.[15] The outcome was favorable in our case despite polyhydraminos.

It has been hypothesized that the obstruction to the lymphatic drainage of sequestration might be responsible for pulmonary effusion and associated pulmonary hypoplasia. Unilateral massive pleural effusion should also be added to the list of unusual forms of pulmonary sequestration in the neonatal period. Most pulmonary sequestration show a spontaneous partial or complete regression during the course of pregnancy and can be managed expectantly with excellent prognosis. However, in some cases there might be increase in size and may develop massive pleural effusions with mediastinal shifting.[16]

Currently, intrafetal vascular laser ablation of the feeding vessel (VLA) is a recommended highly effective treatment option.[17] Those ELS detected postnatal if asymptomatic can either undergo surgical resection or transcatheter arterial embolization or observation without treatment. However, symptomatic ELS should undergo surgical resection. Currently, baby is discharged and has returned to Dang. As the facility of intrafetal vascular laser ablation of the feeding vessel was not available at Patan hospital and other nearby referral centre; it was

TABLE 3 Previous studies with pleural effusion and pulmonary sequestration.

Study	Weeks of gestation during delivery	Vigorous resuscitaton requirement	Polyhydraminos yes/no	Thoracostomy performed	Outcome	Diagnosis (antemortem or postmortem)
Dresler S	34 WOG	Yes	Not commented	No	Expired	Post mortem
Bliek AJ, Mulholland DJ	34 WOG	Yes	Yes	No	Expired	Post mortem
Horowitz RN	30 WOG	Yes	Yes	No	Expired	Post mortem
Lucaya et al.	34 WOG	Yes	Not commented	No	Expired	Post morterm

Abbreviation: WOG, weeks of gestation.

not considered in the reported case. Baby is planned to be evaluated at 3 months for spontaneous regression or the possible need of invasive procedure.

5 | CONCLUSION

It is important for pediatrician to consider pulmonary sequestration as a possible diagnosis in a fetus with evidence of pleural effusion and polyhydramnios. Neonates with pleural effusion may present with respiratory distress and may need immediate aspiration of fluid to relieve lung compression along with mechanical ventilator support. Doppler USG, CT angiography may aid in diagnosis of pulmonary sequestration.

AUTHOR CONTRIBUTIONS

Sajal Twanabasu: Conceptualization; writing – original draft; writing – review and editing. **Shiva Prasad Sharma Chalise:** Writing – review and editing. Both the authors were actively involved in managing the case at neonatal intensive care unit of Patan Academy of Health Sciences.

ACKNOWLEDGMENTS

None.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare.

DATA AVAILABILITY STATEMENT

Data will be provided by the corresponding author upon reasonable request. Images uploaded in the separate files.

CONSENT

Written informed consent was obtained from the parents to publish this report in accordance with the journal's patient consent policy.

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How to cite this article: Twanabasu S, Chalise SPS. Neonatal pleural effusion associated with pulmonary sequestration: A case report. *Clin Case Rep*. 2024;12:e8956. doi:10.1002/ccr3.8956