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Pulmonary sequestration causing severe cardiac failure requiring lobectomy in an extreme preterm infant

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ABSTRACT

We report a case of a large, extralobar pulmonary sequestration in a preterm infant born at 25 weeks gestational age. A computed tomography (CT) angiogram demonstrated that the arterial supply arose from the celiac trunk while an abnormally large, single left pulmonary vein drained the sequestration. This, along with the large patent ductus arteriosus (PDA), created a double left to right shunt, which resulted in severe, high output cardiac failure. Despite aggressive medical management for 3 weeks, he remained critically ill and developed renal failure. Therefore, after multiple, extensive multi-disciplinary discussions with the family, resection was offered as the only possibility for survival. He underwent a left thoracotomy and resection of the extra-lobar sequestration, which was occupying the lower two-thirds of his left hemithorax. To our knowledge, this is the youngest patient in the literature to undergo resection of an extra-lobar sequestration. Management challenges in terms of balancing the cardiac failure against the timing, approach and success of surgical intervention are also discussed along with a review of the literature.

Pulmonary sequestrations are congenital lung malformations consisting of ectopic nonfunctional lung tissue with their own systemic arterial blood supply and without communication with the tracheobronchial tree. It is theorized that an accessory lung bud which develops from the ventral aspect of the primitive foregut leads to formation of the sequestration [1,2]. Sequestrations remain a rare diagnosis, with an estimated incidence of 0.15–1.8% [3].

We present an unusually large, extralobar pulmonary sequestration in an extremely premature neonate, which posed significant hemodynamic and surgical challenges.

1. Case report

A 25-week gestational age male infant was born to a 33-year-old primigravida woman by forceps-assisted vaginal delivery. On antenatal ultrasound at 24 weeks, a large echogenic mass (3.4 cm × 4.5 cm × 4.8 cm) was seen in the left hemithorax displacing the mediastinum to the right (Fig. 1a and b), The Congenital Pulmonary Airway Malformation Volume Ratio (CVR) was calculated as 1.59. The lesion had not been noted on prior antenatal ultrasounds, suggesting rapid growth. There was no evidence of pleural effusion or fetal hydrops.

The mother presented at 24 + 1 weeks with polyhydramnios and preterm labor, and was given 2 doses of betamethasone. Within a few days, the baby was delivered and was immediately intubated for poor respiratory effort. A chest x-ray confirmed a large mass occupying most of the left hemithorax (Fig. 2). Surfactant was administered. APGAR scores were 1, 1 and 3 at 1, 5 and 10 min respectively. His birth weight was 857 g. He was admitted to the Neonatal Intensive Care Unit (NICU). His immediate postnatal course was complicated by respiratory distress syndrome (RDS) and a patent ductus arteriosus (PDA), further compounded by the effects of the chest mass. On day 6, a CT Angiogram of the chest revealed a large pulmonary sequestration occupying the lower two-thirds of the left hemithorax with a grossly abnormal vascular supply: arterial supply from the celiac trunk with venous drainage into an abnormally large single left pulmonary vein (Fig. 3a and b). This, along with the large PDA, created a double left to right shunt,

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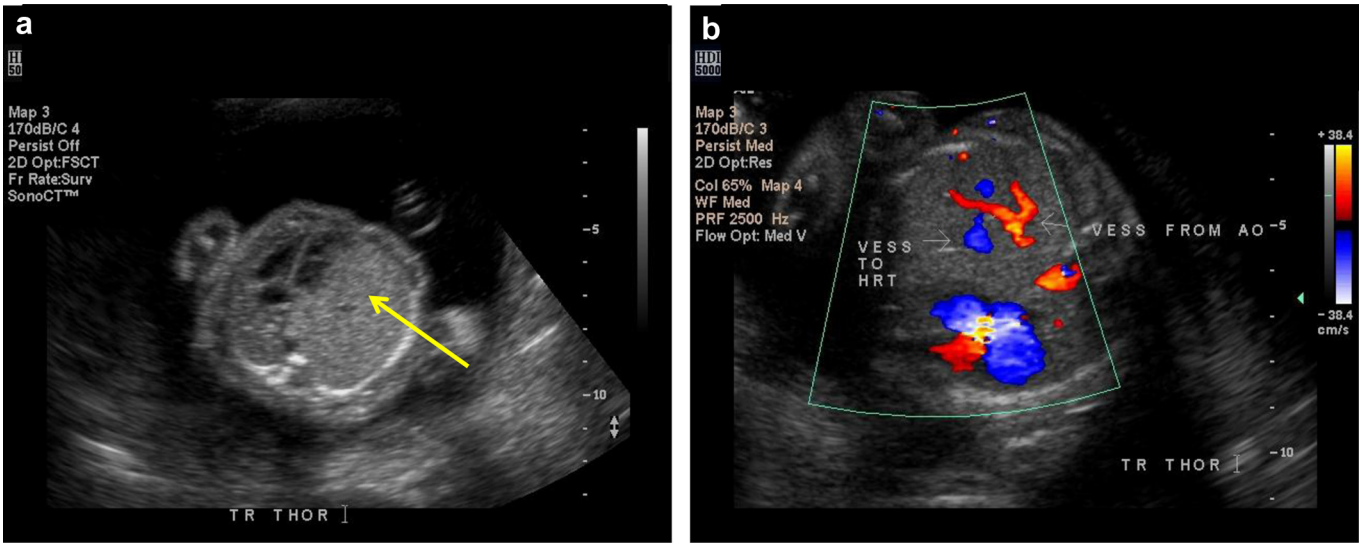

Fig. 1. a and b: Prenatal ultrasound images. The yellow arrow shows the large mass occupying the left hemithorax.

resulting in a high output cardiac failure state with a severely dilated left atrium (LA) and left ventricle (LV) on echocardiogram. Significant fluid restriction, digoxin and lasix were used in the first 3 weeks, along with inotropes and hydrocortisone for diastolic hypotension. Six doses of indomethacin were given on days 2–7 in an attempt to close the PDA. Frequent multi-disciplinary discussions were held with the family, especially with regards to the high mortality risk associated with operative intervention. Unfortunately, his high output cardiac failure persisted and he developed renal insufficiency in the third week, due to the combined steal by the sequestration and the PDA. Mortality seemed likely without intervention. Consideration was given to embolization of the arterial vessel by interventional radiology, but with limited evidence of the success of this approach [4] as well as the very small size of the infant, surgical removal remained the only option. The risks of general anesthesia, bleeding, infection, death and inability to resect the mass and only clip the feeding vessel if the patient was too unstable were all explained to the parents, who consented to the thoracotomy and lobectomy.

On day 23, he underwent a left thoracotomy. Intra-operatively, a large abnormal-appearing mass occupied the lower two-thirds of his chest. There was an obvious feeding artery coming through the diaphragm into the mass. There was also a large pulmonary vein draining the mass. After resection, two normal lobes were visible, suggesting that this was an extra-lobar sequestration. Immediately after resecting the mass, his intraoperative inspiratory pressures decreased from 36 cm H_2O to 22 cm H_2O, while his PEEP decreased from 15 cm H_2O to 10 cm H_2O. He tolerated the surgery well.

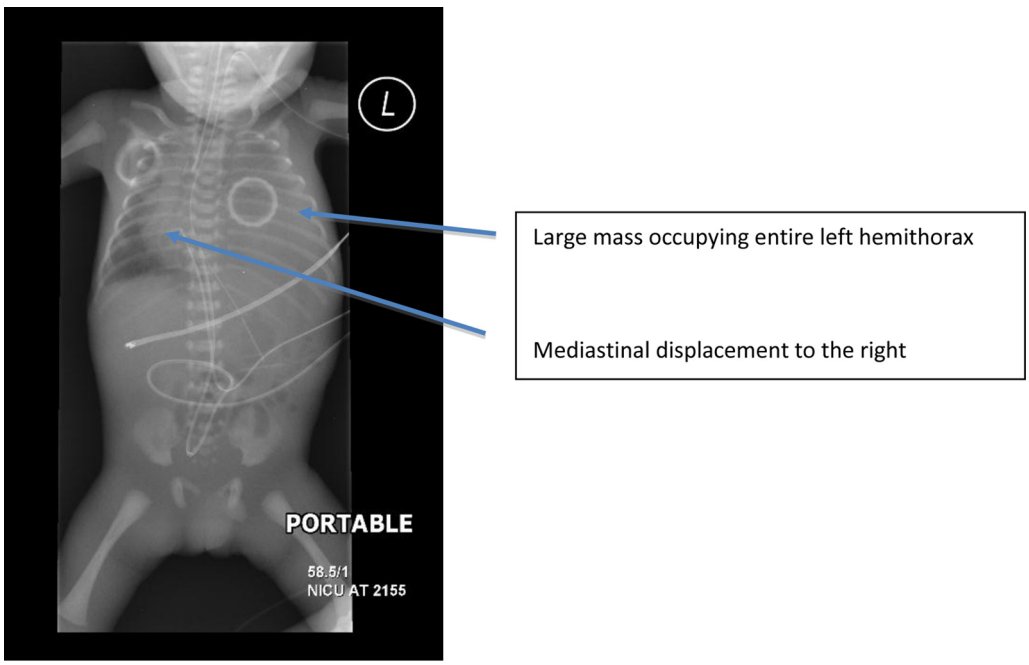

Fig. 2. Immediate postnatal chest x-ray.

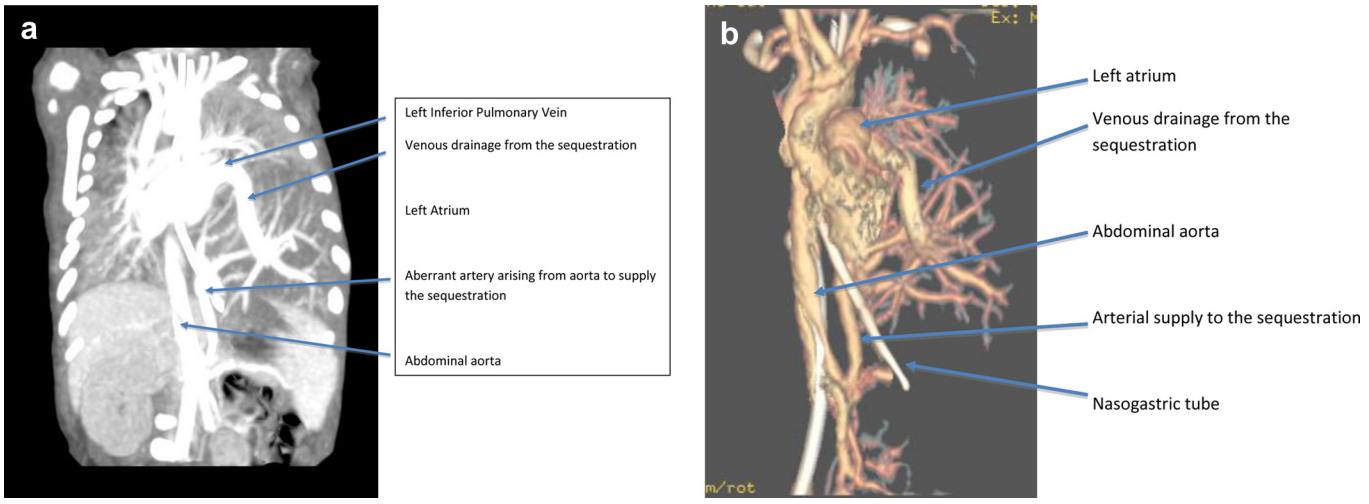

Fig. 3. a: CT angiogram. b: CT angiograph: digital subtration image (3D reconstruction).

Histopathology confirmed the diagnosis of pulmonary sequestration. He was briefly extubated on day 35 before being reintubated for pneumonia, necessitating jet ventilation. After a 7-day course of intravenous dexamethasone, he was finally extubated to nasal cannula positive airway pressure (NCPAP) on day 53. He received intermittent aldactazide for chronic pulmonary edema and chronic lung disease of prematurity. His PDA re-opened post-surgery, but was managed conservatively and eventually closed. Other extreme prematurity complications arose during his stay, such as apnea of prematurity, multiple episodes of feeding intolerance (without necrotizing enterocolitis), need for use of prolonged total parenteral nutrition (TPN), direct hyperbilirubinemia, two episodes of sepsis (with *Bacillus* and *coagulase negative Staph aureus*), bilateral grade II intraventricular hemorrhage (IVH) and stage I–II retinopathy of prematurity (ROP). He was transferred to home hospital on day 112 at a corrected age of 47 weeks. He needed home oxygen until 9 months of age and respirology follow up was delegated to his pediatrician after age 2, since he had no apparent respiratory problems. His current active issues are mild dystonic cerebral palsy and scoliosis. He is currently almost 7 years old and continues to do well from a respiratory standpoint.

2. Discussion

Pulmonary sequestrations can be intralobar or extralobar, based on the absence or presence of investing pleura, respectively. Intralobar sequestrations are encased by normal lung parenchyma, and are more common than extralobar ones. Extralobar sequestrations receive an arterial supply from an aberrant branch of the thoracic or abdominal aorta. Venous drainage is systemic but occasionally occurs into the pulmonary vein [5]. Only 25% of sequestrations are extralobar, and they are more common in males and on the left side. Over half of extralobar sequestrations have associated pulmonary anomalies such as bronchogenic cysts, lung hypoplasia, congenital pulmonary airway malformation (CPAM), or congenital diaphragmatic hernia. Pulmonary sequestration can regress in utero although some continue to grow rapidly and cause pleural effusions, polyhydramnios and/or fetal hydrops [1,6,7]. The Congenital Pulmonary Airway Malformation Volume Ratio (CVR) is used to predict which fetuses are at risk of developing hydrops [8]. At birth, only 25% of sequestrations present with respiratory symptoms. If not detected antenatally, the diagnosis is usually made incidentally on a chest radiograph. Usual presenting features in symptomatic patients are respiratory distress, congestive heart failure (CHF), feeding intolerance in cases of enteric communications and rarely, back pain and hemoptysis [2,3].

Primary diagnostic modalities are Doppler ultrasound, CT and MRI. Doppler ultrasound is useful for demonstrating the typical appearance of the sequestration as well as documenting its vascular supply, but is not ideal for surrounding parenchymal abnormalities. CT angiography has the advantage of demonstrating all of the above. MRI has no radiation risk, but requires a general anesthetic and may be inferior to CT for thin walled cysts and emphysematous pulmonary changes [3]. The natural history of prenatally diagnosed lung masses is variable and is usually managed with maternal transport, planned term delivery in a hospital with Pediatric Surgery and NICU support, evaluation for in utero procedures and postnatal resection if symptomatic or persistent. However, if hydrops develops, there is an almost 100% mortality [8,9].

Possible fetal interventions include: open fetal surgery, sclerotherapy, laser coagulation and/or thoracoamniotic shunting [10]. However, outcomes to date have been poor and the ideal fetal treatment for pulmonary sequestrations is unknown [10,11]. Fortunately, many extralobar pulmonary sequestrations decrease dramatically in size in utero and may not need treatment after birth [11]. Large sequestrations are unusual [7,10].

Standard postnatal treatment usually involves a formal thoracotomy with segmentectomy or lobectomy. Thoracoscopic surgery has also been used in select cases. Interventional radiology uses various embolization techniques for occlusion of feeding arterial vessels. Cho et al. [4] reported the use of trans-arterial embolization in 42 children with a pulmonary sequestration, whose average age at procedure was 17 months. Eighty-three percent showed partial regression, 9.5% had no regression and only 7.1% showed complete regression. In our patient, thoracoscopy and/or embolization were contraindicated due to the large size of the sequestration in an extremely small sized infant.

The unique features of this case were the sheer size of the malformation and the limited options for management in a 25-week preterm infant. Sequestrations have been reported in infants born as preterm as 28–29 weeks [4,9] and presenting with CHF at 32 weeks gestation [12]. There have also been reports of sequestrations in unusual locations: suprarenally adherent to the stomach [13], infradiaphragmatic [14] and with unusual venous drainage to the portal vein [15]. With such varied locations, differential diagnosis for sequestrations should include neuroblastoma [13], mediastinal teratoma, CPAM and diaphragmatic hernia [15]. No data exists for infants born as early as 25 weeks with a symptomatic sequestration.

In our case, the combination of the large arterial branch from the abdominal aorta feeding the extralobar sequestration as well as the left to right PDA shunt, resulted in both high output cardiac failure and renal failure, despite aggressive medical management. This 'steal' phenomenon has been previously described [16]. Resection (or embolization of the feeding vessel) is required to stop this process. Surgery in symptomatic congenital lung malformations is associated with higher perioperative complications, longer operative-time, increased blood loss and post-operative morbidity, with longer lengths of stay and more frequent postoperative complications [17]. Fortunately, our patient tolerated his thoracotomy and resection well and immediately improved clinically. In hindsight, although the CVR was just under 1.6, we postulate that preterm labor may have been lifesaving for this infant, as this large sequestration had already resulted in polyhydramnios and might have resulted in hydrops and/or fetal demise without intervention.

3. Conclusions

Extralobar sequestrations are rarely symptomatic in the neonatal period. CT angiography provides a good imaging modality for diagnosis as well as helps delineate vascular supply to assist surgery. We describe the finding of a large sequestration in a 25-week gestational age newborn, resulting in high output cardiac failure and renal failure. Resection resulted in immediate clinical improvement. The need for resection should be individualized to the nature, size and anticipated clinical course of the lesion.

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