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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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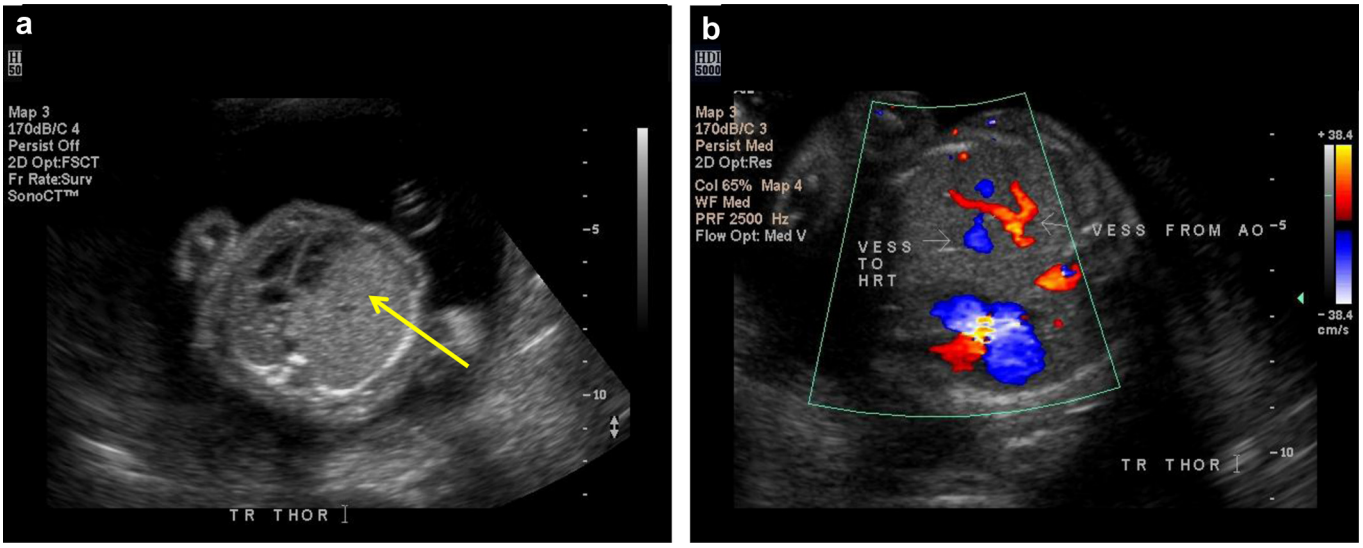

Fig. 1. a and b: Prenatal ultrasound images. The yellow arrow shows the large mass occupying the left hemithorax.

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.

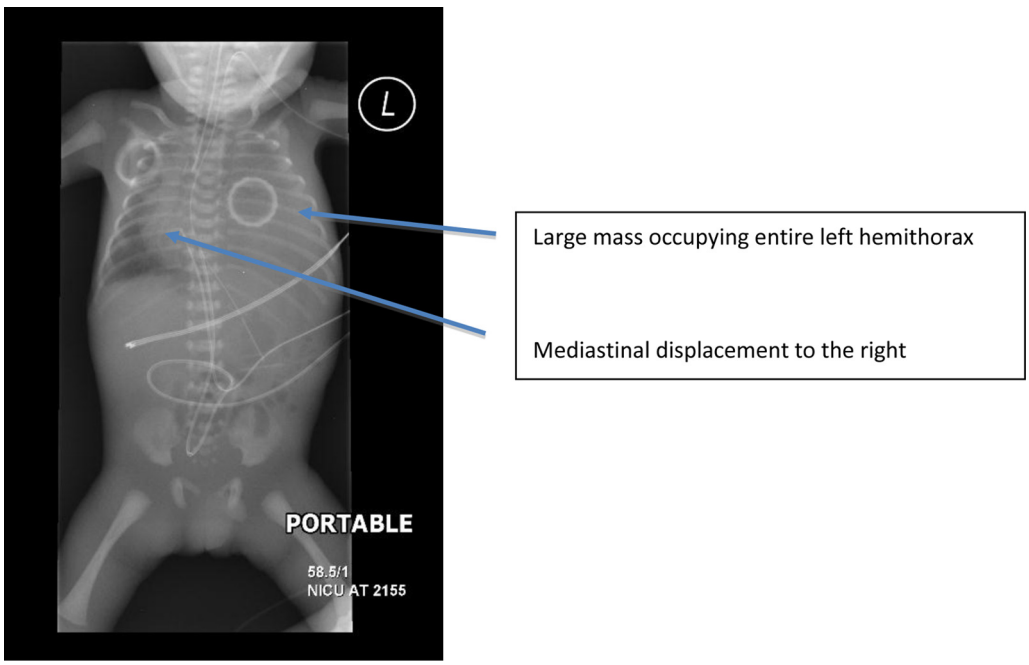

Fig. 2. Immediate postnatal chest x-ray.