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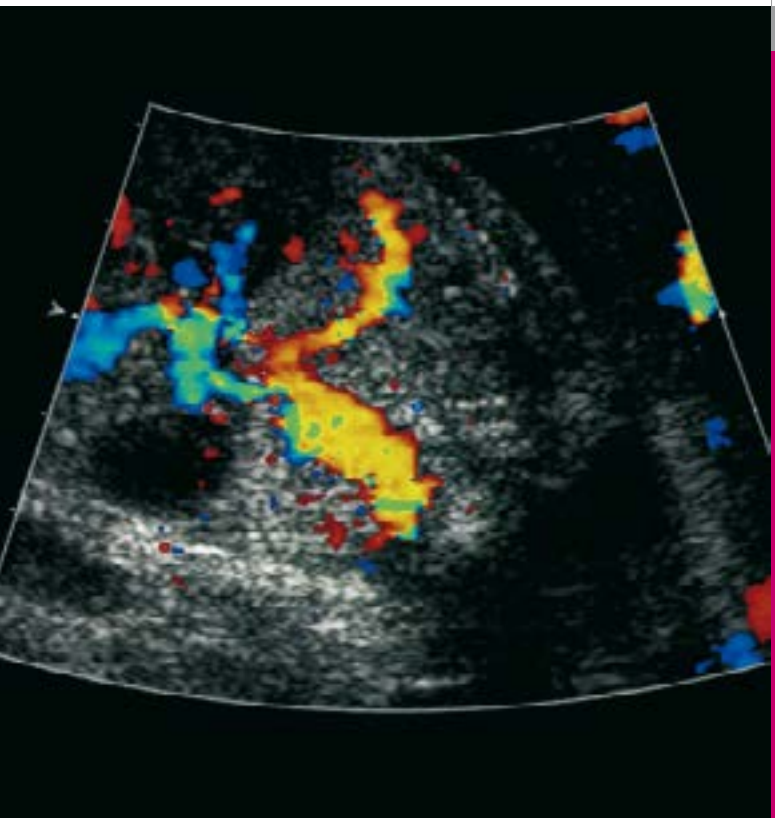
Bilateral pulmonary sequestration in a neonate



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Woerner A, Schwendener K, Casaulta C, Raio L, Wolf R, Zachariou Z, Nelle M, Division of Neonatology, (WA, SK, NM), Department of Pediatric Respiratory Medicine (CC), Division of Pediatric Radiology (WR), Department of Pediatric Surgery, University Children's Hospital, Bern, Switzerland (ZZ), Department of Obstetrics, University Hospital, Bern, Switzerland (RL)

INTRODUCTION

Bronchopulmonary sequestration is a lung malformation that may be detected by antenatal ultrasonography or in the context of recurrent pulmonary infection in children. We present the very rare case of a newborn with bilateral intralobar pulmonary sequestrations. Given the risk of mediastinal shift, pleural effusion or fetal hydrothorax, this malformation required intensive prenatal and postnatal surveillance. Lung function testing gave additional information for optimal postnatal management.

CASE REPORT

Prenatal ultrasonography in a primigravida at 20 weeks of gestation revealed echogenic masses in the right hemithorax (Fig. 1) with systemic arterial supply from the aorta (Fig. 2). Mediastinal shift towards the left side was present. In serial ultrasound examinations, persistence of the lesion was confirmed, with otherwise appropriate fetal development. No pleural effusion or fetal hydrops were noted. A male infant was delivered vaginally at 41 2/7 weeks of gestation, with a birth weight of 3710 g and Apgar scores of 8, 9 and 9 at 1, 5 and 10 minutes, respectively. Arterial and venous umbilical cord pH were 7.27 and 7.36, respectively.

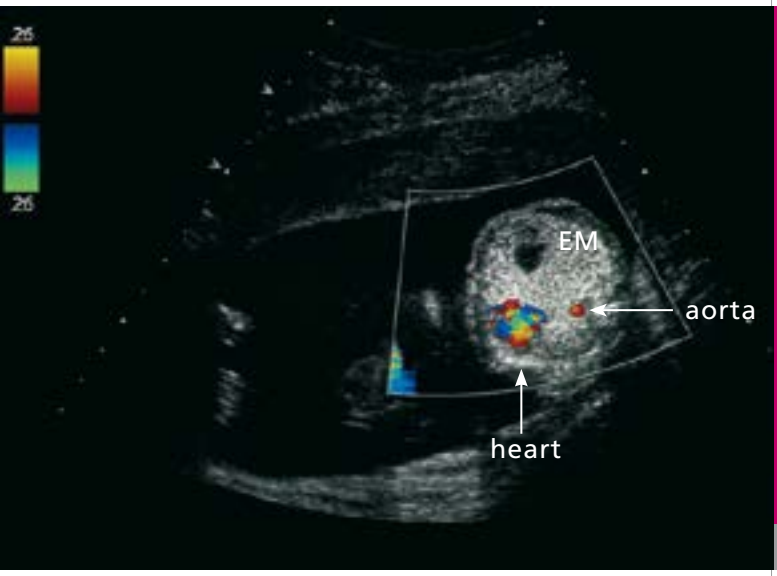

Fig. 1

Prenatal Doppler ultrasonography shows an echogenic mass (EM) in the fetal thorax at 20 weeks of gestation.

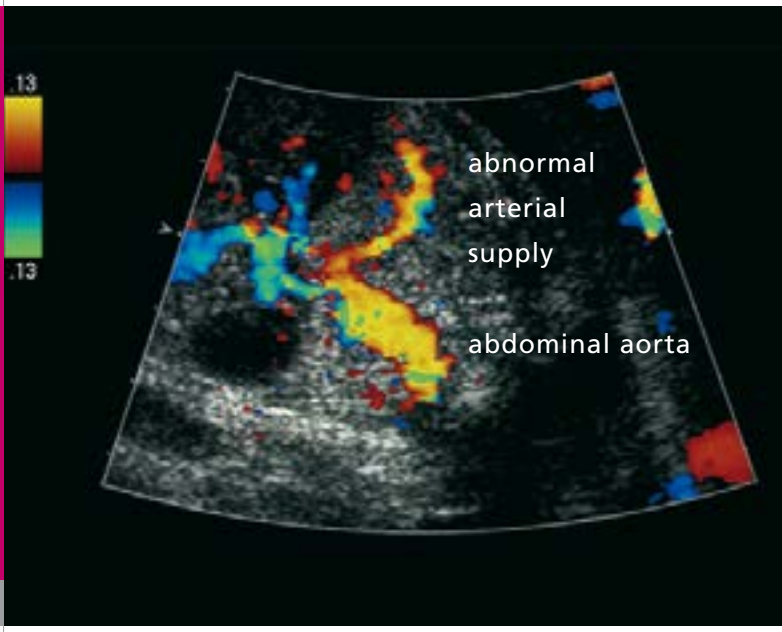

Fig. 2

Prenatal Doppler ultrasonography shows an abnormal arterial supply originating from abdominal aorta.

Apart from isolated intermittent tachypnea between 60 and 90/min, physical examination and transcutaneous oxygen saturation were normal. On day 4, a CT scan confirmed not only a lesion in the right lower lobe measuring 55x32x46 mm but also revealed a second lesion in the left lower lobe measuring 17x26x26 mm (Fig. 3). 2D reconstruction of CT images showed that the arterial supply originated from a single anomalous celiac artery with supradiaphragmatic bifurcation. Venous vessels from both sides appeared to drain into the left atrium (Fig. 4). The echocardiogram demonstrated an otherwise normal anatomy without any signs of volume overload. On the third day of life, the patient developed clinical signs of infection and was treated with amoxicillin and amikacin with rapid recovery. He was discharged home on day 10 of life with persistent intermittent tachypnea of 60-80/min but otherwise unremarkable physical examination.

Further diagnostic work-up included an upper gastrointestinal barium study which showed a normal upper gastrointestinal tract without any evidence of a fistula or extravasation. Baby lung function testing using a multibreath gas washout method was done. The washout pattern was compatible with significant ventilation inhomogeneities, possibly related to a lung structure with multiple functional compartments.

Radiographic follow-up studies done at the age of 2, 3 and 4 months of age showed enlargement of both sequestrations. Preoperative CT angiography confirmed the arterial supply and venous drainage as previously shown. At 7 months of age, bilateral thoracotomies were performed and revealed intralobar sequestrations on both sides. On the right side, the lower lobe was resected, and, on the left side, an atypical resection of segments VII and X was performed after diagnostic thoracoscopy. Pathological and histological examination of the resected tissue confirmed the surgical diagnosis. The postoperative course was uneventful and the infant was discharged home after 8 days. A follow-up chest radiograph three weeks after surgery was consistent with atelectasis of the right middle lobe which was confirmed and reopened during bronchoscopy. At the age of 24 months, the child is doing well and no further pulmonary complications have occurred.

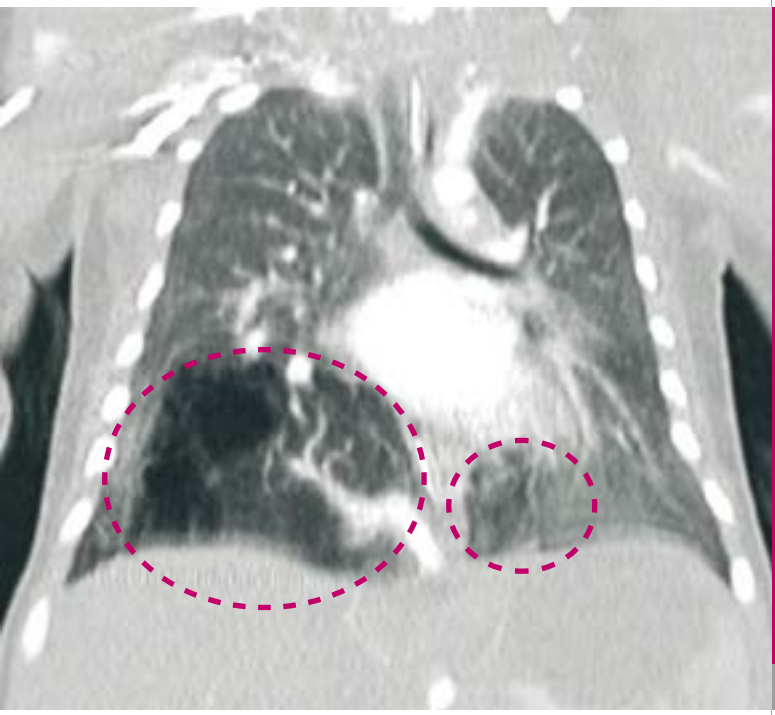

Fig. 3

CT of the chest demonstrates bilateral pulmonary sequestrations in the right and left lower lobes.

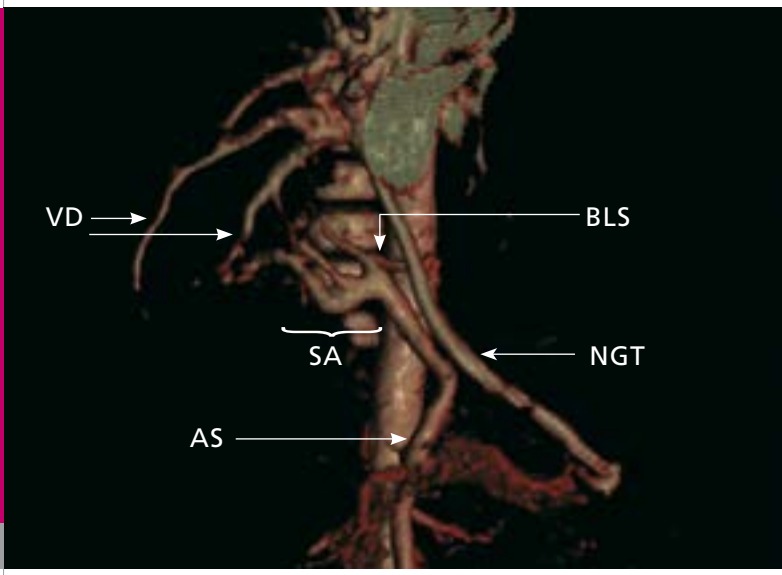

Fig. 4

2D reconstruction of CT images with arterial supply (AS) originating from the celiac trunc with supradiaphragmatic arborisation (SA) and branching to the left sided sequestration (BLS); venous drainage (VD) to the left atrium; nasogastric tube (NGT).

DISCUSSION

Pulmonary sequestration is characterized by the presence of nonfunctioning lung tissue which does not primarily communicate with the tracheobronchial tree and derives its blood supply from the aorta. It can be extra- or intralobar and is usually unilateral, involving the left lower lobe in most cases. Intralobar sequestrations are located within the normal lung tissue and share the same visceral pleura. Extralobar sequestrations are covered by their own pleura and are separated from the normal lung tissue. Secondary bronchial communication is more common in intra- than in extralobar sequestrations, whereas communication with the gastrointestinal tract is seen more often in extralobar sequestrations (1).

Bilateral pulmonary sequestration is a very rare entity, with only few cases reported in the literature. Fifteen cases of bilateral sequestration in childhood have been described in the English literature of whom six with intralobar presentations (2-13). Pulmonary sequestrations are a subgroup of bronchopulmonary foregut malformations which also include congenital cystic adenomatoid malformation, congenital lobar emphysema and bronchogenic cysts. It is postulated that they all have a similar embryological etiology. This is also supported by the fact that in many cases mixed lesions are present (14). The possible mechanism, as proposed by Clements and Warner (15), could be a disruption of the systemic vascularisation of the diverticulating tracheobronchial buds at different times of pulmonary development, resulting in different malformations.

With the improvement of antenatal ultrasonographic screening, prenatal detection of even small thoracic cystic lesions is now possible. In a series of 41 prenatally detected pulmonary sequestrations, Adzick et al. reported that in 28 cases the lesions regressed or even disappeared during serial prenatal ultrasonography (16). The cause for lung lesion regression is unknown. However, fetuses with associated symptoms such as mediastinal shift, tension hydrothorax and non-immune hydrops may require prenatal intervention including fetal thoracocentesis or thoracoamniotic shunt placement. Hydrops may be the result of vena cava obstruction, mediastinal shift and cardiac compression (17). Tension hydrothorax appears to be the result of fluid or lymph secretion from the pulmonary sequestration (18). If untreated, postnatal adaptation and outcome may be complicated and even fatal, e.g. in case of pulmonary hypoplasia due to impaired lung development.

In our case, the large size of the pulmonary sequestration (initially interpreted as unilateral) and the presence of a mediastinal shift to the left side lead to close ultrasonographic follow-up. Fortunately, no tension hydrothorax or non-immune hydrops were noted. However, the size of the lesion remained unchanged. Despite bilateral lung lesions, postnatal adaptation was uncomplicated, consistent with reports of other cases diagnosed postnatally (19-21).

In addition to imaging studies, baby lung function testing can provide important information. Intermittent ventilation of at least one of the sequestrations, suggested by the presence of a multi-compartment washout curve at deep breaths, provided two pieces of information: (1) the likely presence of a secondary connection of one or both sequestrations to the bronchial tree, and (2), as a consequence, an increased risk for complications due to respiratory infection because of the impaired host defense of the sequestrations.

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CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch