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Case Report

Antenatal diagnosis of bronchopulmonary sequestration: A case report and review of the literature ☆,☆☆

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ABSTRACT

Congenital lung malformations are a constellation of pathologies that can be diagnosed antenatally by ultrasound and fetal MRI. Ultrasound is considered the modality of choice for a routine assessment of second-trimester scans worldwide. Bronchopulmonary sequestration (BPS) and congenital pulmonary airway malformation (CPAM) are the 2 most common echogenic chest masses discovered incidentally during routine ultrasound scans in the second trimester. This paper describes BPS and differentiates it from CPAM sonographically in utero. An extensive literature search involving antenatal ultrasound is undertaken to review the most up-to-date understanding of the BPS. Furthermore, a case study at our institution and the literature review will help better describe the salient features of BPS. A 41-year-old female G3P1 visits our department for a routine second-trimester ultrasound. An echogenic lesion with a cystic component is visualized in this scan. Based on the grayscale and color imaging, this complex echogenic lesion was reported as CPAM and was referred to fetal assessment for confirmation. The fetal assessment diagnosed the lesion as BPS because of the pathognomonic feeding vessel from the thoracic aorta. Regardless of the congenital lung mass, any large mass compromising fetal well-being is an indication for intervention. The prognosis of BPS in the absence of fetal hydrops is excellent. A robust collaboration among radiologists, obstetricians, and pediatricians is required for the best outcome for the pregnancy and the neonate.

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Introduction

Bronchopulmonary sequestration (BPS) is a relatively rare pathology discovered incidentally on routine ultrasound scans. It is one of the few congenital lung malformations (CLM), and its incidence has recently increased owing to improved equipment resolution and awareness among sonographers and radiologists [1]. BPS was originally described by Hubber in 1777 as an accessory pulmonary lobe, while Pryce in 1946 coined the term "sequestration" to define the lesion [2–4]. BPS is the second most common CLM diagnosed antenatally, the first being congenital pulmonary adenomatoid malformation (CPAM) [5,6]. BPS is dysplastic or nonfunctioning lung tissue that has no true communication with the anatomical tracheobronchial tree [7]. Furthermore, the dysplastic pulmonary tissue receives anomalous circulation from the thoracic aorta rather than pulmonary vasculature [8]. Additionally, although not pathognomonic, in the case of extralobar sequestration (ELS), the venous drainage is into the pulmonary veins. In contrast, intralobar sequestration is reported to be drained in the azygous system of venous vasculature. There is a subtle disparity in the literature in regards to venous drainage for these 2 pathologies [9,10]. These 3 aspects are some of the prominent distinctive features separating the 2 pathologic diagnoses. The location of the BPS is supradiaphragmatic (85%-90%), subdiaphragmatic (10%-15%), and 90% (left-sided) [11]. BPS is classified into 2 types based on the pleural covering of the nonfunctional lung tissue. Extrapulmonary bronchopulmonary sequestration refers to the type that has a pleural covering separate from the adjacent normal lung and has systemic vasculature. Conversely, intralobar pulmonary sequestration (ILS) is dysplastic lung tissue that is adjacent to the normal pulmonary parenchyma and shares the same pleura with venous drainage into the pulmonary vein [12,13]. Intralobar bronchopulmonary sequestration is thought to form the predominant type of BPS diagnosed in the literature accounting for almost 75% of the cases [9]. Sonographically it is extremely difficult to distinguish intralobar from extralobar types of BPS [14]. CPAM and BPS can coexist in the form of a hybrid lesion. The histology will show characteristics of both the lesions and the cystic lung mass generally receives its vasculature from a systemic vessel in this hybrid BPS [11]. The clinical symptomatology varies depending upon the type of BPS. Hence, diagnosing this pathologic abnormality as early as possible is vital to prevent the patient from recurring symptomatology.

Epidemiology

The incidence of BPS is believed to be around 0.1%-6.4% [7,9,15,16]. Intralobar bronchopulmonary sequestration is 4 times more common than ELS and tends to present itself during adolescence or later years [7]. Intralobar BPS generally presents later in life as chronic cough or fever. It affects both sexes in equal proportions. Extralobar BPS has a male predilection and is usually diagnosed in early childhood [9]. Some authors believe that intralobar bronchopulmonary sequestration might have an acquired etiology rather than a discrete congenital origin [17]. Extralobar BPS is more prevalent in males relative to females, with a reported ratio of 3:1 or 4:1, while intralobar BPS has an equal gender proportion [1,4]

Pathogenesis/etiology

The exact etiology of the condition is not elucidated with certainty. Apart from 1 isolated case report among 2 male offspring, there are no known more extensive studies on genetic predisposition to the development of BPS [14,18]. Some of the leading hypotheses point to events during early embryogenesis that might lead to this pathology. Specific cell adhesion molecules are believed to disrupt the interaction between the mesenchyme and epithelium, leading to pulmonary malformations [19,20]. The role of the HoxB5 gene has also been implicated in the altered expression of crucial transcription factors [21]. Some authors believe that it results from the formation of an accessory lung bud separate from the origin of the normal lung tissue. This accessory lung tissue also acquires distinct vasculature different from the normal pulmonary circulation, thereby remaining independent from the tracheobronchial tree [12]. This pathogenesis supports the development of extralobar BPS [22]. There is ample ambiguity surrounding the origin of intralobar BPS prenatally, and the literature is widely mixed on its pathogenesis. Sequestered lung tissue is mostly nonaerated, but few authors believe that some degree of ventilation might be possible with the normal pulmonary tissue through the pores of Kohn [5]. Intralobar BPS in the postnatal life is known to present as a frequent episode of pneumonia and abscesses [5]. It is for this reason that some authors believe that intralobar BPS variant has an acquired etiology owing to a histological specimen of the inflamed mucosa and the fact that this variant presents later in the life cycle [4,23,24].

Symptomatology

Wei and Li reported in their paper on retrospective analysis of almost 2000 BPS patients that cough or expectoration accounted for most of the clinical symptomatology. This paper placed symptoms like fever, hemoptysis, chest pain, and dyspnea in decreasing order of presentation [3]. Some authors consider pulmonary infections to be the predominant symptom in postnatal life in BPS [25,26]. Repeated pneumonia not responsive to antibiotic treatment might lead to the diagnosis of an underlying congenital lung malformation [10]. Patients may present with respiratory distress and feeding difficulties especially, in the case of the extralobar variant of BPS [9].

Diagnostic modalities

Ultrasound and MRI are the 2 predominant radiologic modalities that can detect BPS antenatally [25,27,28]. The widespread use of ultrasound around the globe and the cost-effectiveness (relative to MRI) associated with this modality make it an indispensable tool for the early diagnosis of BPS in utero. However, fetal MRI serves as an adjunct to antenatal diagnostic imaging to ultrasound. Fetal MRI is reported to characterize

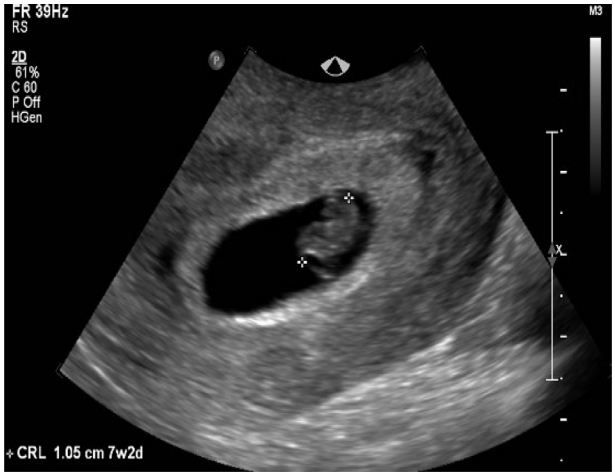


Fig. 1 – The Crown lump length (CRL) corresponds to 7 weeks 2 days.

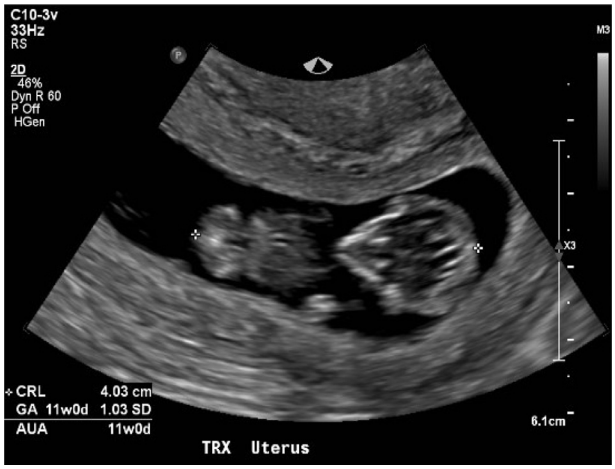


Fig. 2 – The CRL corresponds to 11 weeks 0 day.

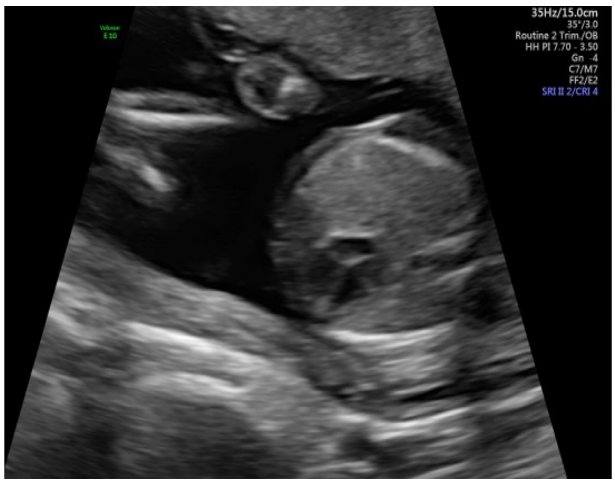


Fig. 3 – No obvious gross pathology is seen in the right chest. Heart is evident on the left in this transverse plane.

the outline of the lesions better and commonly appear as a hyperintense mass on T2 weighted images [1,29]. Some authors consider radiographs the first imaging modality following delivery for any congenital lung lesion detected antenatally [30]. If an abnormality suspicious for a malformation is detected on radiography, CT with contrast is the modality of choice in adults to diagnose BPS, with thin sections (< 1 mm slice thickness) and timed for arterial opacification with as rapid a contrast injection as possible for the age to allow for identification of any feeding vessel [31]. Some authors recommend a CT scan at 3-6 months postnatally, regardless of the radiography or asymptomatic history in the patient [20]. Most authors consider CT scan with contrast as the gold standard to rule out BPS or CPAM in infants [5,29,32,33]. In 1 study, CT is reported to have 100% PPV as compared to 65% for fetal MRI and ultrasound [32].

Differential diagnosis

Some of the most common differential diagnoses to consider for BPS are CPAM, congenital lobar emphysema (CLE), congenital diaphragmatic hernia (CDH), bronchogenic cysts, pulmonary agenesis, pericardial tumors, esophageal duplication, teratoma, congenital high airway obstruction syndrome, neuroblastoma and thymic masses [5,34]. Extralobar BPS should be considered in the differential diagnosis of any intra-abdominal mass with or without a cystic component extending through the diaphragm into the mediastinum [23]. For this reason, congenital diaphragmatic hernia (CDH) is often in the differential for this type of BPS. There are some anomalies reported to be associated with BPS, especially with the extralobar variant [10]. Some of these are Scimitar syndrome, Goldenhar syndrome, Hirschsprung's disease, esophageal atresia with tracheoesophageal fistula, neuroenteric cysts, vertebral anomalies, pulmonary hypoplasia, diaphragmatic eventration, cardiac anomalies, aberrant pancreatic tissue, pectus excavatum, bronchogenic cyst, and pericardial malformation [14]. Most of these differential diagnoses and associated anomalies can be differentiated from BPS with various sonographic techniques. In equivocal cases, fetal MRI might be helpful to separate BPS from the ambiguous differential diagnosis.

Treatment/management (antenatal or postnatal)

Serial ultrasound scans are the best way to assess the size and overall fetal development once the diagnosis is established antenatally [35]. In most cases (as was in this patient), the lung lesion tends to regress or involute and is not visualized in the third trimester or close to the parturition. Hence, close observation is the best advisable strategy for these patients. On the contrary, the presence of fetal hydrops is an indication of intervention [25].

About 75% of the BPS cases tend to involute in utero by term [36]. Postnatal imaging is recommended to rule out the possibility of incomplete resolution of the congenital lung malformation [32]. In some institutions, it is a standard practice to perform a plain chest X-ray on day 1 of the postnatal life with subsequent CT if patients are symptomatic [32]. Radiographs were used in some institutions before CT, probably owing to

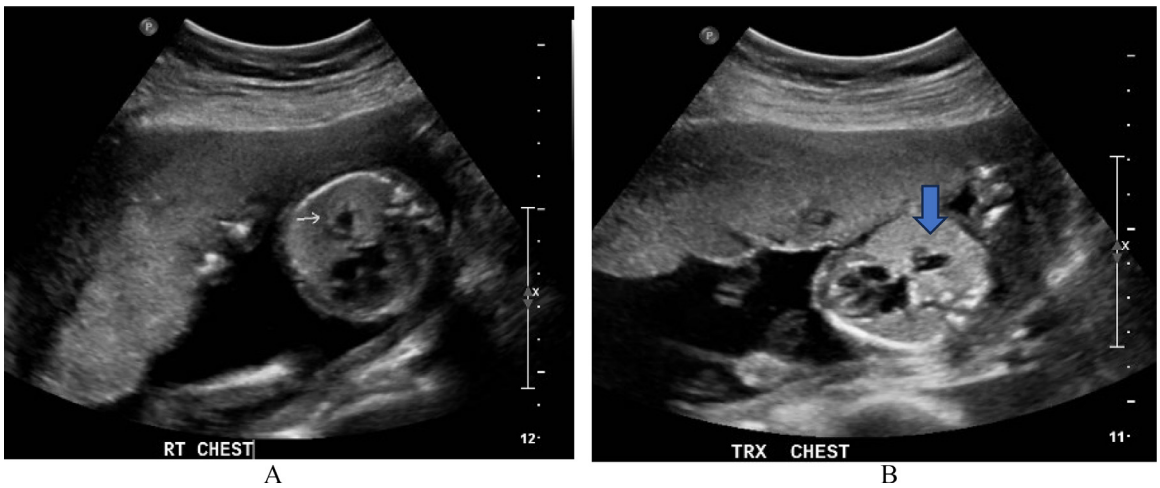


Fig. 4 – (A) A complex echogenic lesion in the right chest with cystic component (arrow). (B) Better optimization of the lesion (arrowhead).

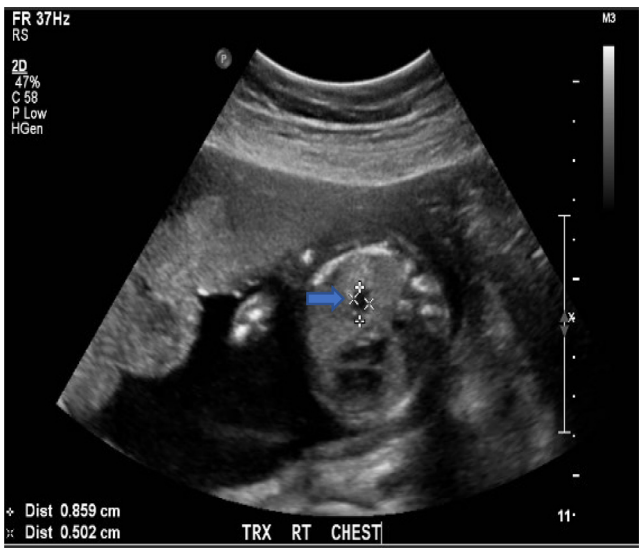


Fig. 5 – The cystic complex in the echogenic lesion measured 8.6 mm in the largest dimension (see arrowhead).

the differences in cost-effectiveness of the 2 modalities. Regardless, Chest CT and X-ray are essential imaging modalities for the postnatal management of BPS or any other congenital lung lesion [37].

Therapeutic intervention is required if complications arise because of the lesion's progressing size and fetal hydrops' development. Maternal steroids, ex utero intrapartum treatment (EXIT), fetal mass resection and thoracoamniotic shunts are known treatments for BPS and other lung lesions. Few authors consider steroid administration a first-line therapy for congenital lung lesions, especially if the congenital pulmonary airway malformation volume ratio (CVR) is > 1.6 [38–40]. It is hypothesized that steroids might trigger the advancement of fetal lung development from the canalicular to the saccular stage [5]. Ex utero intrapartum treatment (EXIT) procedure, fetal lung mass resection and thoracoamniotic shunts are more aggressive therapeutic approaches towards this pathology and implemented only if the disease progresses after the conservative measures (ie, observation and maternal steroids) [41]. EXIT procedure is a highly specialized surgical technique in which the fetus is partially delivered, that is, it is still attached to the placenta and undergoing fetomaternal blood exchange while the surgical team removes the nonfunctioning lung mass. After the removal of this space-occupying lesion, the fetus is delivered completely [42].

Prognosis

The overall prognosis of BPS is reported to be favorable by many authors [6]. Numerous studies have described increasing pathologic lung mass volume/size as the predominant factor that dictates the BPS's prognosis [5]. The use of CVR (CPAM Volume Ratio) has been established in the literature for this purpose. CVR is calculated by measuring the 3-dimensional size of the mass multiplied by 0.52 and dividing the same by the fetal head circumference [43]. Hydrops in the setting of BPS carry a worse prognosis and, without intervention, lead to 100% fetal demise [25,44]. BPS without hydrops is known to have an excellent prognosis, especially if the CVR is less than 1.6. Although CVR as a prognostic tool was initially devised for CPAM, it is also being employed to assess the prognosis of BPS [45]. One study stressed the utilization of CVR before 24 weeks as a better predictor of neonatal outcome relative to CVR performed later in the third trimester [35].

The prognosis of BPS also depends on the presenting symptomatology [46]. For instance, neonatal heart failure, respiratory distress and recurrent infections are some sequelae due to BPS requiring clinicians' emergent attention. Symptomatic intralobar BPS indicates a focal resection of the nonfunctioning tissue, while asymptomatic extralobar BPS is monitored clinically for an impending worse symptomatology. Some authors recommend arterial embolization of the sequestering

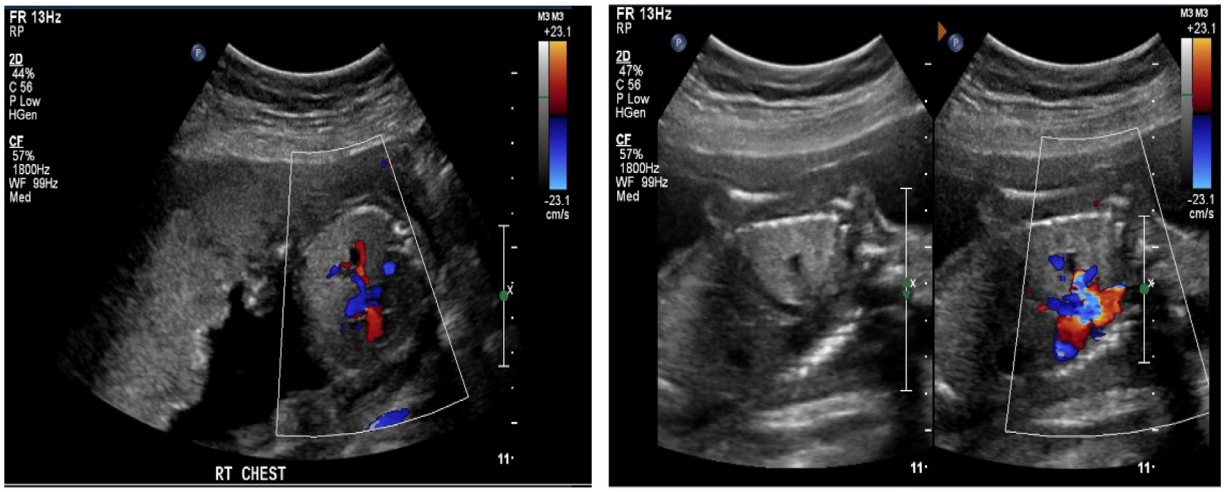


Fig. 6 – (A) Transverse: no flow in the cystic. (B) Sagittal: no flow in the cystic component of the echogenic lesion.

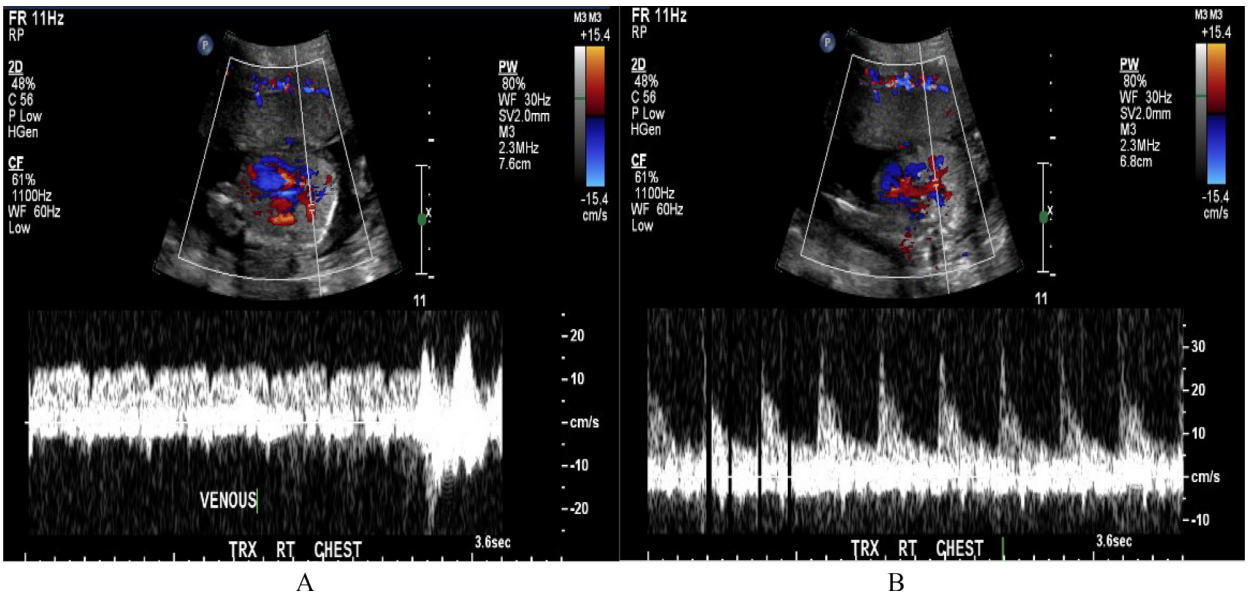


Fig. 7 – (A) Transverse: venous flow. (B) Transverse section: arterial flow.

feeding vessel as an alternative to resection by an interventional radiologist or a cardiologist [1,4]. Overall, the typical progression of BPS and other CLMs ranges from 100% regression by term to fetal death in utero [21].

Case report

A 41-year-old female G3P1 presents to our department for a first trimester dating ultrasound. A healthy first trimester was reported with gestational age at 7 weeks 2 days, almost concordant with the last menstrual period (LMP) at 7 weeks 4 days (Fig. 1). The fetal heart rate is recorded at 154 beats per minute.

The patient was referred for the second time to our department to rule out threatened abortion due to bleeding and declining "beta HcG" hormone levels. No cause of vaginal spotting or decreasing "beta HcG" levels is described in the current exam. The pregnancy was extrapolated from the last exam and is reported as concordant with the current exam (Fig. 2).

Since the patient was above 35 years of age, it is a routine practice in obstetrics in our institution to recommend maternal serum screen. In this patient, the maternal serum screen returned positive with a ratio of 1:209 for trisomy 18 or Edwards syndrome. Thus, the patient was referred to the fetal assessment unit at 17 weeks. The scan was reported normal, and no fetal chest abnormality was detected (Fig. 3).

Our institution performs a routine obstetrical ultrasound exam at 20 weeks. Hence, the patient was scheduled for an expected second-trimester obstetrical exam at 20 weeks gestation in our department. During this sonographic examination, the fetus biometry measurement placed the pregnancy

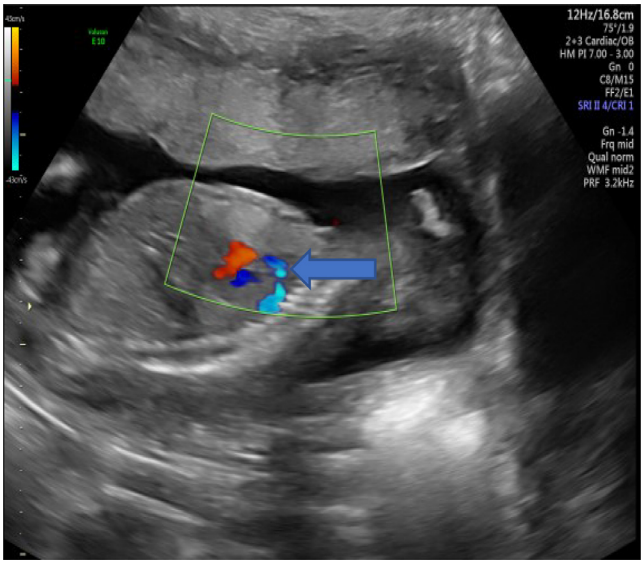


Fig. 8 – Vascular pedicle off the thoracic aorta supplying the echogenic lesion (see arrow).

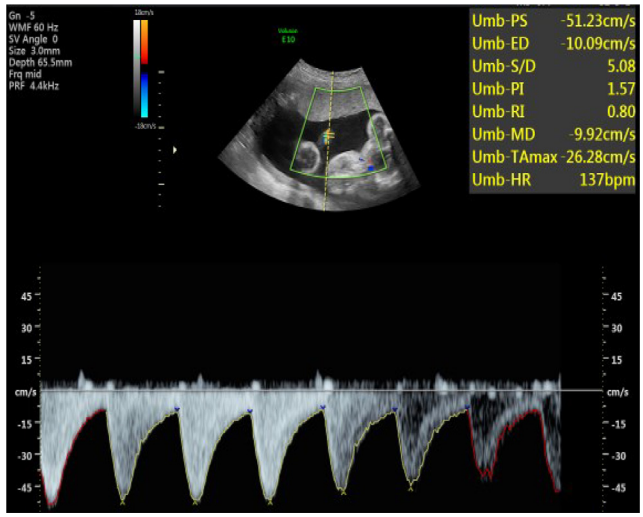


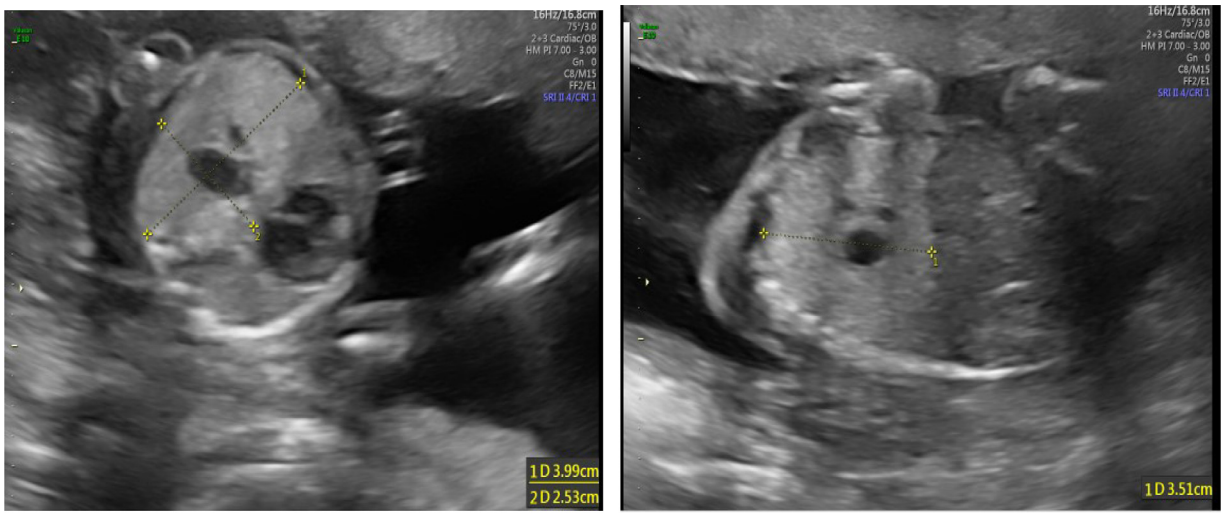
Fig. 10 – Fetal assessment scan showing S/D ratio=5.08.

at 20 weeks 5 days. For the first time, the sonographer noticed a complex echogenic lesion in the right chest with a cystic component within (Figs. 4A and B). From a clinical perspective, the 4-chamber view is ideal for ruling out fetal cardiac abnormalities, but it also works very well for ruling out fetal chest anomalies [10]. An inexperienced sonographer may only use these 4 chambers to rule out cardiac pathology and may not focus discretely on the adjacent structures, thereby missing the potential congenital lung malformations, like the BPS.

There was no flow in the 8.6 mm cystic component (Figs. 5, 6A and B), while the echogenic lesion had both venous and arterial flow (Figs. 7A and B). The sonographer could not discreetly establish the origin of this flow. No evidence of pleural effusion, hydrops, ascites, mediastinal shift of the thoracic organs or additional fetal structural anomalies were visualized and reported in this scan. A preliminary diagnosis of CPAM was made by the sonologist based on the cystic component in the echogenic lesion, and the patient was referred to fetal assessment for further expert opinion and management purposes.

The first fetal assessment ultrasound is performed a week later, and a formal diagnosis of bronchopulmonary sequestration is established based on the feeding vessel from the aorta (Fig. 8).

Moreover, the dimension of this echogenic lesion seems to have progressed marginally as compared to the ultrasound performed just a week ago. The current maximum extent of this lesion was about 4 cm in the transverse section and 3.5 cm in the sagittal plane. (Figs. 9A and B).



A B

Fig. 9 – (A) Transverse section of the chest. (B) Sagital section of the chest.

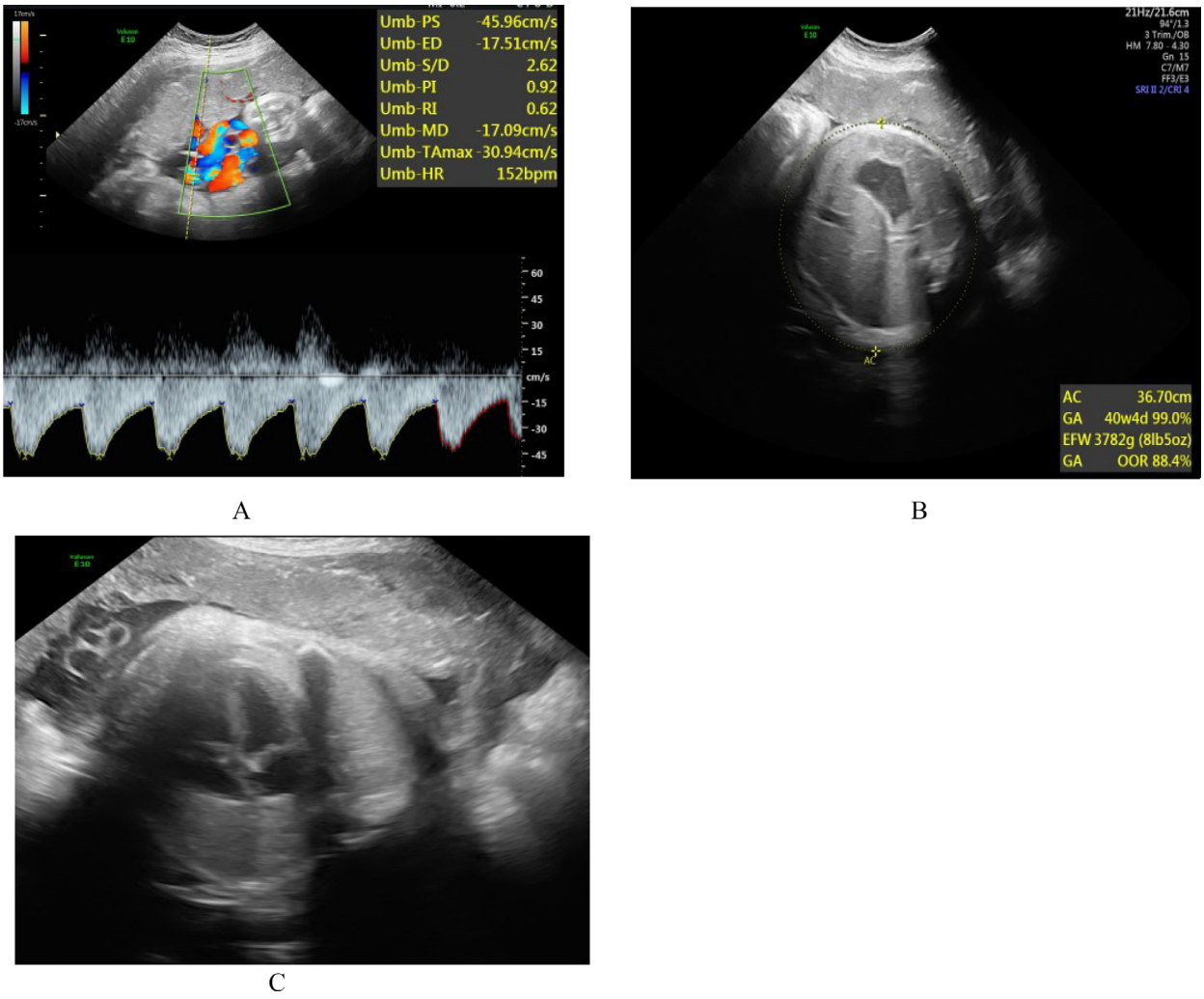


Fig. 11 – (A) Fetal assessment scan in third-trimester. (B) No evidence of echogenic lesion in RT showing S/D ratio 2.62 chest. (C) Third trimester: No gross pathology is seen in the right chest in this image. The heart is along the normal left axis orientation.

The CPAM volume ratio (CVR) for this fetus was around 1.0 without any evidence of hydrops or any other grave symptomatology. Therefore, serial ultrasound scans were planned for this patient in the fetal assessment unit for management purposes. A month later, the fourth serial ultrasound scan revealed an increase in the S/D (systolic/diastolic) ratio in the umbilical artery (ie, 5.08). (Fig. 10) Placental bed is a low resistance fetomaternal circulation system. As the pregnancy progresses, the diastolic flow in the uterine artery decreases, thereby decreasing the S/D ratio in the late third trimester [47]. Close to the parturition, the 50th percentile for the S/D ratio in the umbilical artery is reported to be around 2.18 [47]. Some authors believe the mean S/D ratio during the 40th week is 2.61+/-0.450 [48].

During the late third trimester, this S/D ratio was borderline normal (Fig. 11A), measuring just over 2.6, and the echogenic lesion in the right chest seemed to have regressed or not visualized on the current ultrasound exam. (Figs. 11B and C)

Except for observation, no other intervention is utilized for management purposes in this patient. The neonate had a chest X-ray on day 1 following birth (Fig. 12). The radiologist reported central lucency in the mid-right lung field and suspected incomplete resolution of BPS. Following this report, transthoracic echo (TTE) is recommended for this neonate. The result of this scan revealed no abnormalities.

No follow-up chest X-ray or CT with contrast is scheduled for this infant since the last imaging. Per the most current records, the infant is nonsymptomatic and has no development issues. We would have anticipated a CT with contrast in this infant especially when the prior radiograph (Fig. 12) raised the possibility of a nonresolving congenital lung malformation. We speculate that the CT with contrast was not ordered due to the nonsymptomatic nature of the case.

Discussion

Ultrasound profoundly impacts the diagnosis of CLMs, especially during the antenatal fetal development period. With

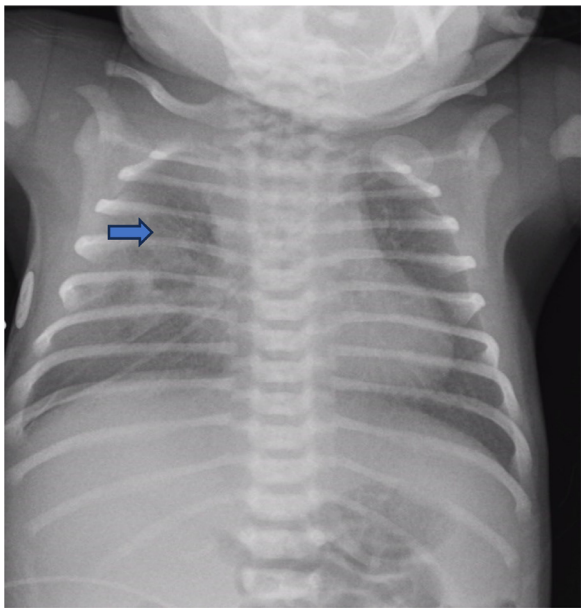


Fig. 12 – A subtle central lucency in the mid-RT chest is visualized in this radiograph.

modern ultrasound equipment, the incidence of CLM, especially CPAM and BPS, has brought sonography to the forefront, particularly during antenatal life. CPAM and BPS are the 2 most common CLMs diagnosed in utero in experienced hands. Aside from the uncommon exceptions, for all clinical purposes, the sonographic diagnosis of BPS is made by finding the pathognomonic feeding vessel from the aorta. For CPAM, the feeding vessel to the cystic mass is primarily from the pulmonary arteries rather than the systemic vasculature [49]. The radiologist should be particularly wary of not relying on the cystic component in the mass for separating CPAM from BPS. BPS presents as an echogenic lesion on prenatal ultrasound and may or may not have a cystic component.

Diagnosing and differentiating the various forms of CLMs is vital since the symptomology, prognosis, and treatment are different. For instance, any development of fetal hydrops in bronchopulmonary sequestration, diagnosed by ultrasound, allows obstetricians to plan pregnancy management, including counselling [32]. However, ultrasound is not helpful in delineating histology of the various CLM lesions. Some authors also believe that prenatal ultrasound is not sensitive enough to depict the pathognomonic feeding vessel to these lesions. Therefore, antenatal MRI or postnatal contrast CT is needed to manage the patient outcome better [1,50].

Any recurrent lung infections with all tests normal during childhood, adolescence or in later life should make the physician think about the underlying congenital lung lesions. Undiagnosed ILS (Intralobar sequestration) has been associated with frequent episodes or recurrent lung infections in later years. Furthermore, BPS is reported in the literature to be associated with lung cancer [51–54]. Some articles have also reported the sudden increase of tumor markers, namely, carbohydrate antigen CA 19-9 and carcinogenic embryonic antigen (CEA), in addition to metaplasia in the bronchial epithelium [54,55]. Hence, diagnosing BPS at the earliest to prevent the patient from undergoing multiple imaging and other tests is necessary.

In some cases, such as in this patient, the previously diagnosed BPS in the second-trimester ultrasound seems to regress by the end of the third trimester [1]. Some authors believe this is due to the isoechogenicity of the lung malformation with the adjacent pulmonary tissue particularly close to the term rather than a true involution or resolution [5,56,57]. In such patients, postnatal CT is highly recommended to rule out the complete regression of the pathology, as mentioned above [45]. Unfortunately, this was not done in this case for some unknown reasons, even though the postnatal CXR raised suspicion of an incomplete lesion resolution.

Conclusion

Diagnostic imaging professionals must detect or rule out bronchopulmonary sequestration, preferably antenatally. Any missed diagnosis might lead the patient to have recurrent pulmonary infections and other respiratory issues later in life. Fetuses diagnosed with BPS antenatally should be appropriately followed up for the resolution/involution of the lesion after birth. The role of ultrasound and MRI in diagnosing BPS or any other CPAM is vital in the in-utero environment. CT with contrast is the modality of choice in the ex-uterine setting, while CXR is also utilized for any congenital lung malformation diagnosed antenatally. All 4 modalities, namely, ultrasound, fetal MRI, Chest X-ray, and CT with contrast, add value to managing these patients during and after parturition. Therefore, close collaboration among radiologists, obstetricians, pediatricians, and other allied healthcare professionals is precious for the best pregnancy outcome.

Author contributions

Gurinder Dhanju: Conception and design of the study and drafting/revising the article critically. Ashraf Goubran: drafting/revising the article critically for intellectual content. Iain Kirkpatrick: revising the article critically and final approval of the manuscript in detail. Sheldon Wiebe: revising the article critically and final approval of the manuscript. Jordan Fogel: revising the article and final approval of the manuscript.

Patient consent

Written informed consent was obtained from the patient on July 17, 2023, for the purpose of this case study. Case Study received formal approval from the Research Ethics and compliance committee (affiliated with the University of Manitoba) for publication, Dated: September 29, 2023. Ethics Reference Number: HS26149 (H2023:270).

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