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Case Report

Pulmonary Sequestration and Congenital Pulmonary Airway Malformation (CPAM) 1 Presented as A Left Suprarenal Mass: A Case Report

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Abstract

Introduction

Pulmonary sequestration is a rare condition accounting for 0.1% to 6% of all congenital lung malformations. The correlation with Congenital Pulmonary Airway Malformation (CPAM) type 1 has not been adequately elucidated.

Case presentation

From routine fetal sonographic screening, a 20-day-old female baby's ultrasound revealed hydronephrosis with enlargement above the left kidney. Magnetic resonance imaging during a follow-up appointment uncovered a left suprarenal lesion of approximately 30 mm, which was not suspected to be adrenal neuroblastoma. After resection, histological analysis was consistent with pulmonary sequestration and confirmed the connection with CPAM type 1.

Conclusion

Surgery remains the only effective therapeutic choice for intra-abdominal pulmonary sequestration. The definitive diagnosis is confirmed through histological examination.

1. Introduction

Congenital cystic lung lesions (CCLL) represent a heterogeneous group of uncommon clinical-pathological anomalies [1]. Congenital Pulmonary Airway Malformation (CPAM) and Pulmonary Sequestration (PS) are the most frequently prenatally diagnosed CCLL [1]. Pulmonary sequestration is characterized by a segment of nonfunctioning pulmonary tissue that does not communicate with the tracheobronchial tree and derives its blood supply directly from

the systemic arterial tree, with an incidence of 0.1–6% among all CCLL cases [2].

There are two distinct subtypes of PS: intralobar sequestration (ILS), which is situated within the visceral pleura of a normal lung lobe alongside functional lung parenchyma, and extralobar sequestration (ELS), which is located outside the functional lung lobe either within the thoracic cavity or beyond it; ELS possesses its own visceral pleura [1]. Furthermore, hybrid lesions combining components from two distinct malformations may also exist [3]. The association of PS with CPAM type 1 has not been thoroughly elucidated and described in genuine literature [1, 4].

The purpose of this paper is to describe a case of type 1 CPAM with a rare combination of extrapulmonary abnormalities discovered in a 20-day-old female patient.

2. Case Presentation

2.1. Patient information and Clinical findings

A 20-day-old female baby, born via cesarean section (CS) from a G3P3 mother, was brought to the clinic by her parents, weighing 3.5 kg. She was referred by a pediatrician for a checkup and presented with no symptoms. During routine fetal sonographic screening, she was found to have intrauterine hydronephrosis and enlargement above the left kidney. Apart from these findings, the baby appeared well and had no other abnormalities.

2.2. Diagnostic assessment

Several hematological and biochemical tests yielded normal results, except for a slightly elevated serum potassium level (5.49mmol/L) above the reference range (3.5-5.1mmol/L). The preoperative level of vanillylmandelic acid (VMA) was within the normal range. A 30 mm mass was detected above the left kidney in sonographic findings, along with an enlargement of the left adrenal gland measuring 21 × 20 × 22 mm, suggestive of adrenal hemorrhage. The baby was placed under observation and conservative follow-up. Subsequently, magnetic resonance imaging (MRI) revealed a left suprarenal lesion of approximately 30 mm, which did not raise specific suspicion of adrenal neuroblastoma. Computed tomography (CT) scan findings also revealed a left suprarenal lesion (Figure 1.), with the mass measuring about 5 × 4 × 3 cm. Levels of vanillylmandelic acid (VMA) in urine and alpha-fetoprotein (AFP) were both within the normal range.

2.3. Therapeutic intervention

Under general anesthesia, laparotomy was performed through a left upper abdominal transverse incision, identifying a lesion above the left kidney that did not involve the adrenal gland. The lesion was situated on the posterior abdominal musculature and was excised completely, with no harm to surrounding vessels and organs. The postoperative period was uneventful, and the child was discharged in good health. The excised mass underwent histopathological analysis, which revealed a disorganized hamartomatous glandular-like structure connected by ciliated cuboidal and columnar bronchial-like epithelial cells (figure 2). The features were consistent with pulmonary sequestration, and the presence of cartilage confirmed an association with CPAM type 1.

2.4. Follow-up

the post-operative period was uneventful. The patient was free from recurrence one year after the operation.

4. Discussion

Congenital cystic adenomatoid malformation, congenital lobar emphysema/congenital alveolar overinflation, polyalveolar lobe, solitary bronchial atresia, PS, and related hybrid lesions, bronchogenic cysts, are common CCLLs. Most of them might not be distinguishable based on clinical impression or radiological study. Numerous imaging-based diagnoses have to be reevaluated after pathological analysis [1]. Routine antenatal obstetric ultrasound discovers a small subset of abnormalities, including fetal abdominal tumors (5%). Although the prenatal discovery of a tumor can lead to treatment dilemmas, the majority of fetal abdominal tumors are benign, allowing the adoption of a wait-and-see approach without changing the time and mode of delivery. The differential diagnosis of intra- and/or retroperitoneal tumors depends on the display of distinctive sonographic features because the fetus lacks clinical signs and symptoms. Only a small number of specific lesions can be examined by the radiologist, and a few infants will need other diagnostic techniques (CT scan, MRI) to provide more relevant information [5].

Pulmonary sequestration is an uncommon congenital abnormality with an unknown etiology. Pryce first recognized the disease in 1946 [6]. It appears in 1:20,000–35,000 individuals and accounts for 0.15–6.4% of all congenital pulmonary abnormalities [1]. Using obstetric ultrasonography during the prenatal period, it can be diagnosed from the 16th week of gestation [7]. The majority of cases of ELS occur in the thorax (90%), mediastinum, pericardium, and within or below the diaphragm. Extralobar sequestration (ELS), particularly in adults, poses a significant diagnostic challenge. Twenty-five percent of cases are discovered prenatally by 18-19 weeks of gestation. Sixty percent of cases are identified by 3 months of age, which is frequently diagnosed in the left suprarenal region, as in the present case. Due to its location, a differential diagnosis including teratoma, hemolymphangioma, adrenal hematoma, and neuroblastoma has to be ruled out [3, 8].

Congenital pulmonary airway malformation was previously named Congenital cystic adenomatoid malformation (CCAM) by Ch'in and Tang in 1949 [9]. During clinical communication, the terminology and classifications might be very confusing for clinicians and pathologists. As cystic change is not always evident, the term CPAM, which has gained broad acceptance, has replaced the previous term CCAM [10]. Congenital pulmonary airway malformation is a hamartomatous mass of disorganized lung tissues with varying degrees of cystic

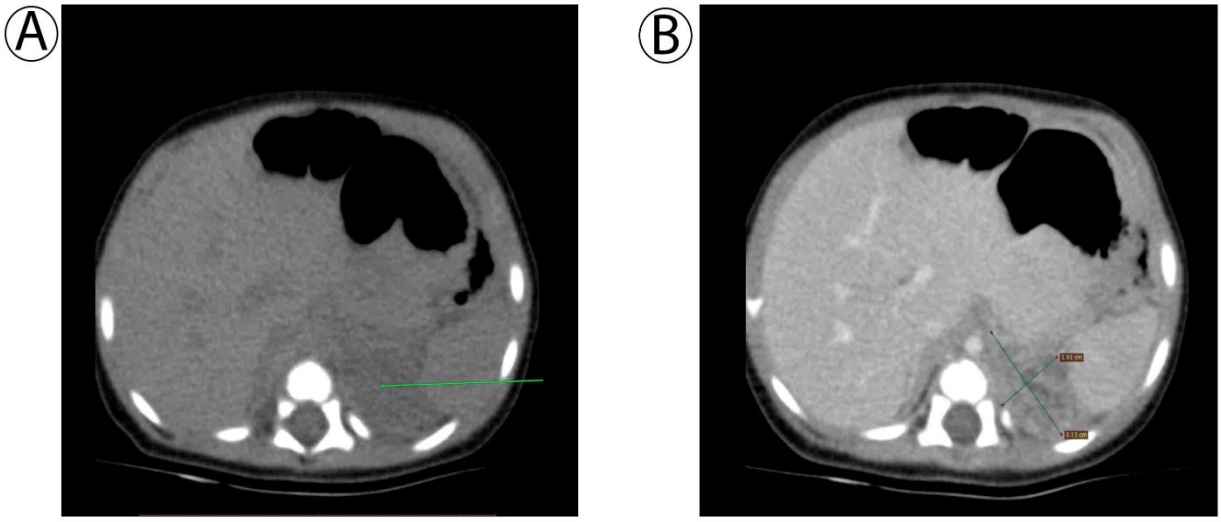

Figure 1. CT scan (A pre, B post contrast) show left side retroperitoneal mass above left kidney, beneath left crura. Homogeneous density of 31x29x19 mm (AP xCC xRL). Showing inhomogeneous enhancement with contrast by focal area of low fluid density

changes, resulting in the formation of many cysts with gaseous or liquid contents that vary in size and distribution [7, 11]. According to the clinical and pathologic characteristics of their 35 cases of CPAM, in 1977, Stocker et al. classified them into three types [12]. In addition to the original types 1, 2, and 3, they subsequently added types 0 and 4, categorizing them as type 0 (tracheobronchial); type 1, bronchial/bronchiolar; type 2, bronchiolar; type 3, bronchiolar/alveolar duct; and type 4, distal acinar [13]. The modified Stocker classification (5 subtypes) has

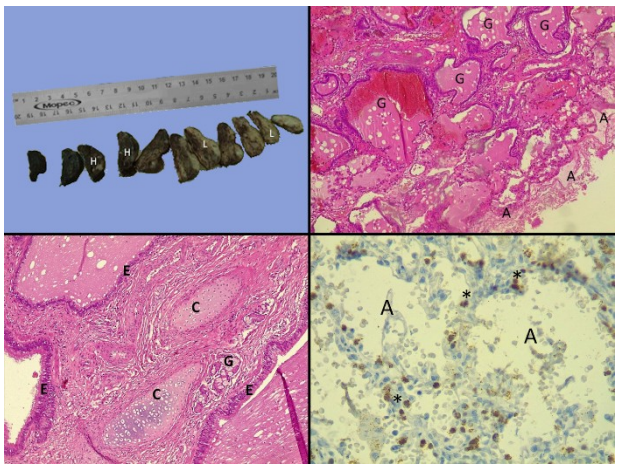

Figure 2. The top left photo shows the gross appearance of the mass, with a spongy, grey-tan texture resembling that of lung (L) and hemorrhagic areas (H). The top right photo shows the admixture of glands lined by pseudostratified ciliated columnar epithelium (G) and alveolar spaces at the periphery (A). The bottom left photo shows a higher magnification view of the glandular spaces (E), hyaline cartilage (C), and seromucinous glands (G). The bottom right photo shows the alveolar spaces (A) with strong, nuclear reactivity of the lining cells (*) for thyroid transcription factor-1 (TTF-1). [Top left: Gross; top right: Hematoxylin and eosin, 40x magnification; bottom left: Hematoxylin and eosin, 100x magnification; bottom right: TTF-1 antibody with diaminobenzidine chromogen, 400x magnification.

been frequently used in practice and literature reports, but its basic pathogenetic mechanism has encountered challenges [13]. In the samples of these cystic lung lesions in prenatal cases, Langston and other subsequent case series studies have discovered bronchial atresia [14, 15]. According to Langston's "bronchial atresia sequence," the degree of bronchial obstruction, its level, and its timing would result in a different pattern of abnormalities. Langston labeled these lesions in a more detailed manner. In Langston's "classification," the terms large cyst type and small cyst type/bronchial atresia correspond to CPAM Stocker types 1 and 2, respectively. In Stocker's defining case series, the CPAM type 1 lesion represented 60%–70% of all cases. CPAM type 2 is the most common type of CCLL, according to Wang's study [1, 16].

After the terminology was developed in 1997, the term "hybrid lesion" has been commonly used to describe PS associated with CPAM in the literature, particularly in clinical and radiological articles [1]. Extra-lobar sequestration has been associated with CPAM type 2, cardiac, chest wall, diaphragmatic, and other pulmonary anomalies. The majority of the reported cases of hybrid PS lesions in children and adults were associated with CPAM type 2 and focused on unusual clinical and radiological features. The PS related to CPAM type 1 has not been completely illustrated and discussed [1-3, 17]. In the present case, a 20-day-old infant had an unusual extrapulmonary sequestration hybrid with type one CPAM.

Routine ultrasound examination in the second trimester is a sensitive method for CPAM prenatal detection. Also, maternal serum tests that evaluate the risk for Down syndrome can occasionally result in CPAM prenatal diagnosis. Pham et al. reported a case in which a regular prenatal checkup during the second trimester revealed a low maternal serum AFP level, resulting in the diagnosis of CPAM in their case [11]. Intra-abdominal PS is commonly located like a sharply circumscribed mass between the diaphragm and kidney in the upper left quadrant, separated from the normal adrenal gland. Prior to excision surgery, a proper diagnosis may be assisted by

knowledge of the characteristic appearance in different imaging modalities. The sonographic appearance of a PS is characterized by a distinctively high level of echogenicity. This phenomenon results from multiple tissue interfaces presented to the ultrasound beam. It has been revealed that the periphery is surrounded by a thin, highly echogenic rim. A systemic feeding artery arising from the thoracic or abdominal aorta may be visualized with Doppler sonography, confirming the correct diagnosis [4]. In the current case, she had an intrauterine ultrasonography scan that revealed a 30 mm mass over the left kidney and an enlarged left adrenal gland with adrenal hemorrhage. Angiogram-based identification of the vascular supply to the lesion may be helpful in preoperative planning, but it is not commonly recommended as CT scans or MRI frequently provide sufficient information. However, a histological study is necessary for a definitive diagnosis [2, 5, 11].

Possible complications of PS include hemorrhagic pleural effusions, malignant transformation, hemoptysis, and the emergence of secondary infections like pneumonitis. Furthermore, torsion with hemorrhage, necrosis, and sepsis are reported in ELS, particularly when the diagnosis is delayed; such cases frequently present as acute surgical emergencies [2]. Intra-abdominal PS generally has a good prognosis and a low risk of infection before surgery. The prognosis may worsen with the presence of other related anomalies, particularly pulmonary hypoplasia. Pulmonary sequestration can undergo malignant transformation; however, the occurrence of a malignant lung tumor in bronchopulmonary sequestration is extremely rare and is frequently associated with ILS. Surgical excision is the only method that provides histological confirmation due to the ELS suprarenal location and the potential for differential diagnosis.

On the other hand, long-term follow-up accompanied by imaging is recommended for an expectant approach in asymptomatic lesions due to the possibility of malignant changes when cystic components are present. The laparoscopic method allows extensive exploration of the abdominal cavity via small incisions, and the magnification allows for good systemic vascular control. Therefore, it is concluded that the risks of surgical morbidity are less significant than the chances of long-term sequestration-related problems [8]. A laparoscopic procedure was performed in the current case, and the postoperative period was uneventful.

5. Conclusion

Hybrid lesions that combine components from two distinct malformations may exist. Due to the initial confusion with neuroblastoma, surgical treatment of potential intra-abdominal PS is justified. The histopathological examination confirms the definitive diagnosis.

Declarations

Conflicts of interest: The author(s) have no conflicts of interest to disclose.

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