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A rare case report of simultaneous occurrence of a pediatric pleuropulmonary blastoma and an intralobar pulmonary sequestration ☆

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ABSTRACT

We are presenting a rare case with the simultaneous occurrence of pleuropulmonary blastoma and an intra lobar pulmonary sequestration. Although there have been cases reported previously with pleuropulmonary blastoma associated with congenital pulmonary malformations, the association with an intra lobar pulmonary sequestration is very rare. The patient, a female, 6-month-old child arrived at our pediatric service with the clinic of cough, respiratory distress, and fever after being treated for 2 weeks for left lung bronchopneumonia according clinical signs and radiographic description but without clinical improvements. Contrast enhanced CT images showed the simultaneous presence of 2 different lesions in the left lung, a heterogeneous mass in the superior lobe without delineation with mediastinal structure compatible with a pleuropulmonary blastoma and a consolidation in the inferior lobe with bronchogram present and a systemic vessel feeding compatible with an intra lobar pulmonary sequestration, both confirmed by histologic examinations after the surgical intervention. Although it is very rare, the simultaneous presence of these distinct embryogenic lesions may occur and radiologist should be aware as the imaging diagnosis may be very helpful for the further management of the patient.

Introduction

Pleuropulmonary blastoma (PPB) is the most common primary malignancy of the lung in pediatric age, an aggressive embryonal neoplasm of the pulmonary mesenchyme which was proposed to be a distinct entity in 1988 [1,2]. Pleuropulmonary blastomas are divided into 3 types: type I PPB are purely cystic tumors with better prognosis and which progress to more aggressive types II and III [3]. Type II pleuropulmonary tumors are mixed tumors with cystic and solid appearance while type III pleuropulmonary tumors are purely solid. Not all cystic type

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I PPB progress to the more malignant types and these cystic cases are classified as type I regressed (type Ir) [4]. Signs and symptoms vary according to the different subtypes. The typical clinic sign of type I tumors is respiratory distress due to air-filled cysts associated or not with pneumothorax while the most frequent clinical signs of types II and III tumors are dyspnea, fever, and chest pain [4,5,6].

On the other hand, bronchopulmonary sequestration (BPS) is a rare tumor described firstly from Pryce in 1946 [7]. It is part of the group of congenital pulmonary airway malformations (CPAM). CPAM are a group of non–malignant developmental anomalies which include a varied group of disorders like congenital cystic adenomatoid malformation, bronchopulmonary sequestration, congenital lobar emphysema, bronchogenic cyst etc. CPAM are relatively rare, with a reported incidence between 1:25000 and 1:35000, while BPS account for 0.15%˜6.4% of all congenital pulmonary malformations [8].

The main feature of this disease is that partial lung tissues separate from the main lung during the embryonic period, grow to generate a cystic mass without lung function and receive its blood supply from systemic circulation arteries, mainly from thoracic aorta and abdominal aorta. Its pathogenesis is not clear and is categorized into congenital and acquired BPS. Acquired theory was proposed by Pryce but an increasing number of studies support the congenital theory which explains that BPS is formed by the growth of the primitive foregut ventral side lung bud during embryonic development period [9,10].

BPS is usually classified as either intra lobar sequestration (ILS) which is located within a lung lobe and shares the visceral pleura with the corresponding lung lobe or extra lobar sequestration (ELS) which is a separate mass of lung parenchyma enclosed entirely by a separate pleural envelope. Intra lobar sequestrations account for 75% of pulmonary sequestrations, being more frequent within the lower lobe (98%) and more often in the left lung (55%) while extra lobar sequestrations constitute the remaining 25% and are typically found between diaphragm and the lower lobe [11].

We are reporting a rare case with the occurrence of these 2 distinct lesions simultaneously; a pleuropulmonary blastoma associated with an intra lobar pulmonary sequestration.

Case report

A female patient, 6-month-old child was presented at the Emergency Pediatric Department of University Hospital Center “Mother Teresa” with the clinic of cough, respiratory distress, and fever. The patient was treated for a period of 2 weeks for a left lung pulmonary bronchopneumonia according clinical signs and radiographic description. The Chest X-ray showed a consolidation in the inferior lobe of left lung with fine bronchogram present and also a small consolidation in the superior lobe of left lung. Contrast-enhanced CT images revealed a heterogeneous lesion in the superior lobe of left lung without delineation with mediastinal structure and minimal left second foramina dilatation (Fig. 1). The radiological diagnosis of this lesion was in favor of pleuropulmonary blastoma, confirmed later by the histopathologic examination after surgical intervention.

CT images revealed also the presence of a consolidation in the inferior lobe of left lung with the presence of bronchogram. The systemic vessel feeding this mass was an accessory branch of thoracic aorta (Fig. 2). The lung consolidation was compatible with pulmonary sequestration confirmed also by the histopathologic examination after surgery.

Discussion

Pleuropulmonary blastoma and pulmonary sequestration represent 2 distinct entities with different prognosis. Although there are case reports of suspected malignant degeneration of CPAM (the group of which BPS account) into sarcomas or other tumor types, in general CPAM and PPB are now regarded as separate entities [12,13,14]. In general, the most frequent reported radiographic features associated with a high likelihood of PPB are heterogeneous low attenuation mass, pleural effusion, contralateral mediastinal shift, and lack of chest wall invasion [15]. In children the lesions which may be confused are type I purely cystic PPB with one of several congenital cystic lesions of the lungs part of congenital pulmonary airway malformations (CPAM) and in these cases a careful pathologic examination is required [5,12,16]. In our case the nature of lesion, a heterogeneous mass, without delineation with mediastinal structure and the presence of minimal left second foramina dilatation were strong radiological features suggesting the diagnosis of PPB type II. In fact, prior to 12 months’ of age, type I PPB lesions are more frequent while type II and III lesions are rarely seen [16] as in our case.

The importance of making the differential diagnosis between PPB and CCAM including pulmonary sequestrations is the high grade of malignancy of the PPB lesions especially type II and III which require surgical resection and subsequent chemotherapy [17]. Also, it is well known that PPB are part of DICER-1 gene syndrome where germline mutations of this gene are associated with increased risk of a wide variety of neoplastic conditions, including pleuropulmonary blastoma, cystic nephroma, nasal chondromesenchymal hamartoma, ovarian Sertoli-Leydig cell tumors, pituitary blastoma, nodular thyroid hyperplasia or thyroid carcinoma etc. Patients should be evaluated very carefully to exclude the presence of a concomitant pathology and surveillance strategies are recommended too [4,18]. So pediatric patients with cystic lung masses should be the subject of DICER1 germline testing especially when the appearance of cysts is with septa, multiple, bilateral, or identified in infancy. In addition, patients who are asymptomatic and being considered for a non–operative management strategy of CPAM should undergo DICER1 testing to exclude a PPB lesion [19].

The second lesion in the inferior lobe of left lung was compatible with a bronchopulmonary sequestration type ILS. The cystic lesions in ILS type may communicate with normal

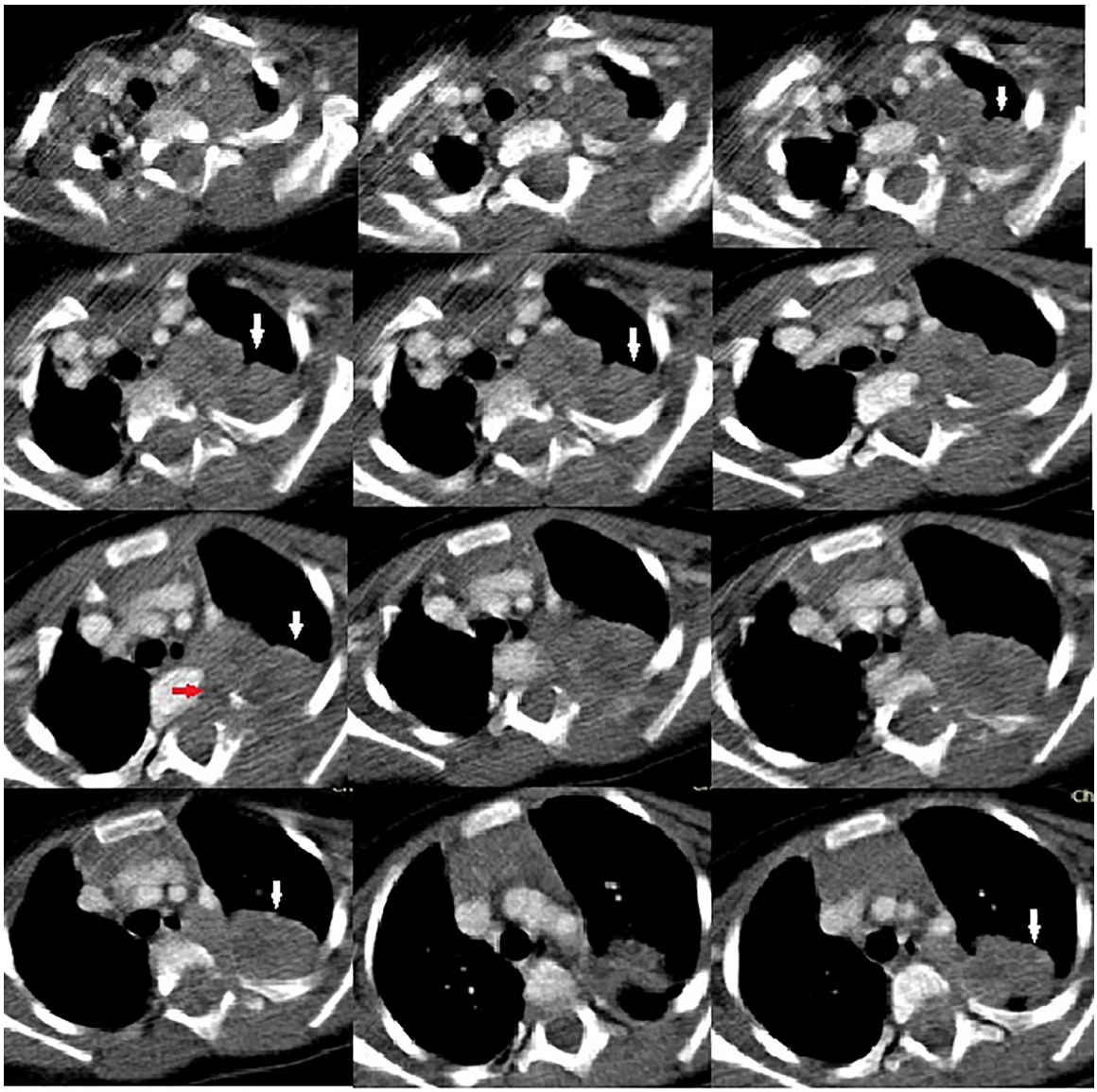

Fig. 1 – Contrast-enhanced CT images show a lesion in the superior lobe of left lung compatible with pleuropulmonary blastoma (white arrow) and minimal left second foramina dilatation (red arrow) (Color version of the figure is available online.)

bronchus which easily leads to repeated infection and produce the clinical manifestations of fever, cough, expectoration, cyanosis as in our case while in ELS type the sequestration lung is outside the lung lobe and its cystic lesions do not communicate with normal bronchial tree so it may be asymptomatic for a long time but in infancy may produce severe acute respiratory distress syndrome [20,21].

Radiographic features strongly associated with pulmonary sequestration include any area of pulmonary hyperinflation and the presence of a systemic feeding vessel which arise mostly from the thoracic aorta (75%) as in our case or abdominal aorta (15%-20%) [22].

Our patient clinical symptoms were cough, respiratory distress, and fever. Patients with similar clinic and ILS type lesion in imagery are often misdiagnosed with pneumonia, lung abscess, pulmonary tumor, and should be evaluated carefully during imagery examinations.

Regarding the treatment of pulmonary sequestration, surgery is preferred to avoid the recurrent infections, pulmonary fibrosis or hemoptysis and careful preoperative identification of systemic arterial supply and venous drainage from a pulmonary sequestration is necessary to avoid hemorrhage during surgery [23]. In our case due to the clinic of the patients and the presence of another lesion of malignant nature like PPB type II, the surgical intervention was the only option of treatment.

According to a large systematic review which assessed the simultaneously occurrence of PPB and CPAM, pleuropul-

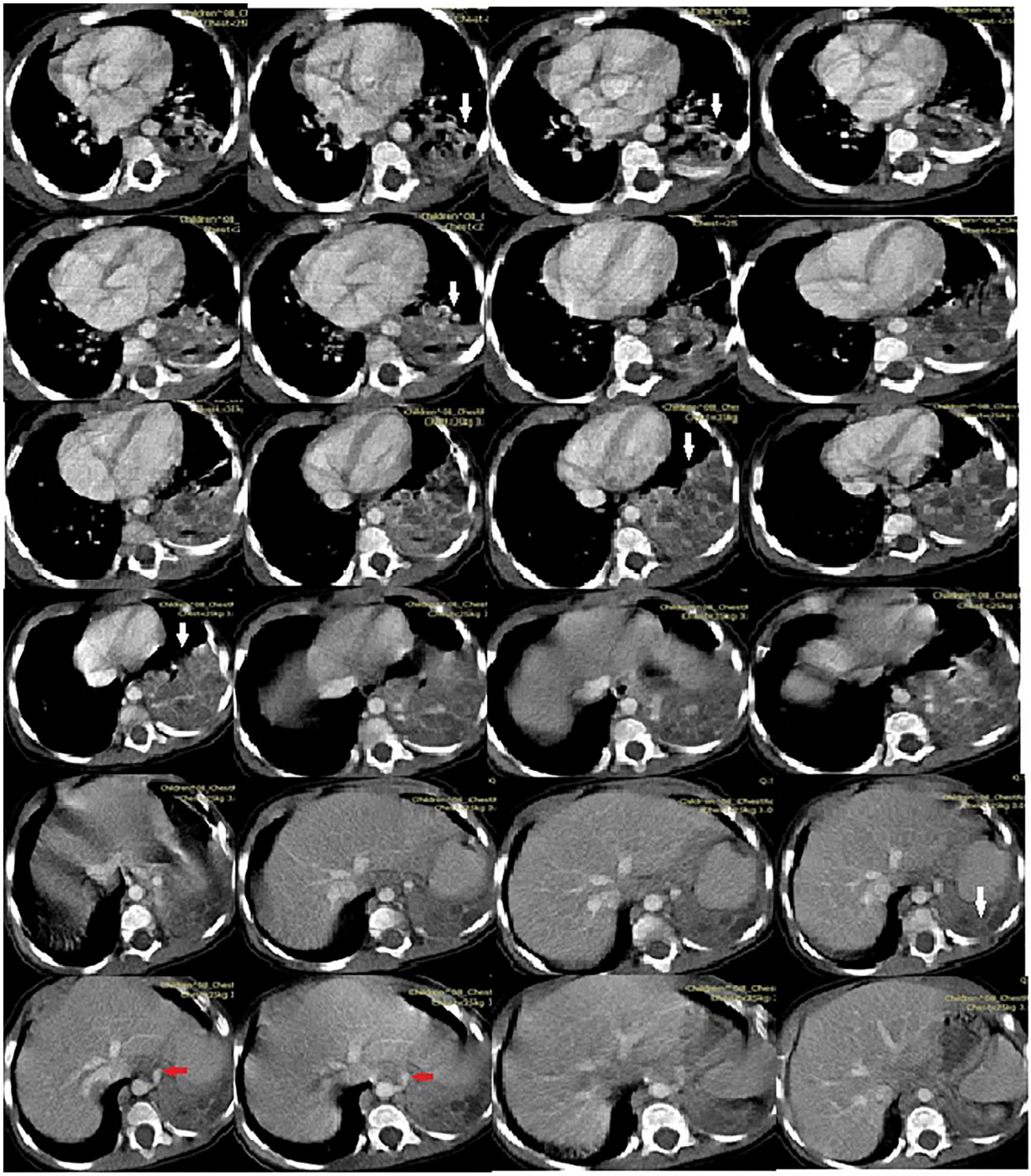

Fig. 2 – Contrast-enhanced CT images show a lesion in the inferior lobe of the left lung compatible with a pulmonary sequestration (white arrow); the systemic vessel feeding the lesion is an accessory branch of thoracic aorta (red arrow) (Color version of the figure is available online.)

monary blastoma has been reported most frequently with congenital cystic adenomatoid malformation (CCAM), CPAM type IV, bronchogenic cyst, congenital lobar emphysema and extra lobar type of sequestration (ELS) while the association between PPB and ILS type is rare [24].

Conclusion

The simultaneous presence of pleuropulmonary blastoma with pulmonary sequestration is very rare especially with ILS

type. In addition to clinical evaluation imaging examinations are very important in the correct diagnosis of these rare embryogenic chest tumors and very helpful for the future management of the patient.

Patient consent

The personal details of patient included in the article has been removed before submission.

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