Pleuropulmonary blastoma in extrapulmonary lung tissue: A case report

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A B S T R A C T
We report a case of pleuropulmonary blastoma (PPB) occurring in a very unusual location, extrapulmonary lung tissue, as possible extralobar pulmonary sequestration. The patient was a girl aged two years and six months with respiratory symptoms. Infection of a congenital pulmonary malformation was suspected based on computed tomography findings. The operative findings revealed a partly adherent mass located between the upper and middle right lobes, and a feeding vessel. Histological analysis showed the mass to be PPB. A small portion of normal-looking pulmonary tissues was confirmed in the subcapsular area of the mass, suggesting that the tumor originated in extrapulmonary lung tissue. The patient underwent postoperative chemotherapy. No recurrence or metastasis of the PPB has occurred during the 10-year follow-up. When considering the treatment options for an extralobar pulmonary lesion, the possibility of PPB should be considered, although its occurrence may be extremely rare.

Pleuropulmonary blastoma (PPB) is a rare intrathoracic tumor of early childhood. It has often been described as associated with cystic lung diseases, such as congenital adenomatoid malformation (CCAM) [1–3]. However, its association with extralobar lung tissue has not been reported in the literature. Here, we report a girl with repeated respiratory infections, whose initial diagnosis was suspected CCAM. However, a histological analysis showed her to have PPB in extrapulmonary lung tissue, possibly within an extrapulmonary sequestration (EPS).

1. Case report

A girl aged two years and six months presented at hospital with cough and fever. A chest radiogram showed right pulmonary consolidation. The CT findings were suggestive of infection of a CCAM and the patient was referred to our institute for further evaluation and treatment. She was febrile at presentation, with reduced respiratory sounds in the right thorax and an elevated white blood cell count. Her medical and family histories were unremarkable. She was treated with antibiotics. A follow-up CT obtained a month later, after her general condition had improved, showed a well-defined, large cystic mass with a solid component in the right thorax. A right thoracotomy was performed. The right lung showed incomplete lobulation between the upper/middle and middle/lower lobes. The mass was located between the upper and middle lobes and appeared to be covered by its own pleura. It was clearly separated from the pulmonary lobes, although there was slight adhesion to the upper lobe. There was no connection to the trachea or gastrointestinal tract. A feeding vessel was confirmed and resected, and the mass was completely removed. The mass measured 80 × 60 × 60 mm and weighed 155 g. The cut section revealed cystic and solid lesions with hemorrhage (Fig. 1). The solid lesions were fragile and nodular in appearance. Microscopically, the tumor was completely covered by a fibrocollagenous capsule and showed diffuse growth of neoplastic cells, with normal-looking ciliated epithelial linings in some areas (Fig. 2A). Some neoplastic cells showed differentiation toward striated muscle. Neoplastic cartilage and hemorrhagic necrosis were also noted. Immunohistochemically, the neoplastic cells were positive for vimentin, desmin, muscle actin, focally positive for myoglobin, and negative for cytokeratin. The epithelial linings were positive for cytokeratin and thyroid tran-
Small amounts of normal-looking pulmonary tissues were noted between the fibrocollagenous capsule and the tumor (Fig. 2B), suggesting that the tumor arose from extrapulmonary lung tissue. Careful review of the preoperative CT revealed what appeared to be an intercostal artery as the feeding artery (Fig. 3); however, histological examination proved this to be a muscular-type artery. The postoperative CT showed no defect or distortion of the right lung, and there was no disruption of the distal bronchial tree. The final diagnosis was PPB arising in extralobar pulmonary tissue. The patient was discharged after completing postoperative chemotherapy. There has been no recurrence or metastasis of the PPB during 10 years of follow-up.

2. Discussion

Pulmonary sequestration is an uncommon congenital anomaly, characterized by an anomalous systemic blood supply and dysplastic lung tissues that have no connection to the tracheobronchial tree. It is classified into intralobar and extralobar types, the former contained within the normal lung and the latter separated from the normal pulmonary lobe, outside the visceral pleura. EPS is less common than its intralobar counterpart, and is more often associated with congenital anomalies such as congenital diaphragmatic hernia, lung and chest wall deformities, foregut malformations, bronchogenic cysts, and complex cardiac anomalies [4–6].

Interpreting the origin of PPB in our patient was very difficult because the artery confirmed during surgery was a muscular type and may have been a parasitized vessel. However, the resected tumor was an independent mass existing outside the normal pulmonary lobes, with no connection to the gastrointestinal tract. It was covered by fibrocollagenous tissue, separate from the visceral pleura that outlined the normal lung lobes. There was no pedicle-like feature that would have suggested a tumor protruding from a normal pulmonary lobe. Moreover, the postoperative CT scan showed no defect or distortion of the right lung tissue, and there was no disruption of the distal bronchial tree. Based on these data, the tumor was considered to have developed from extralobar pulmonary tissue, possibly sequestered lung. Because the growth of the tumor was expansive, the elastic-type feeding artery may have become indistinguishable, although there was no way to prove this.

The management of pulmonary sequestration is controversial, especially in EPS. Patients with intralobar pulmonary sequestration (IPS) often develop respiratory symptoms, most commonly respiratory-tract infections and recurrent pneumonia. There have been very few reports of patients with IPS who remained asymptomatic throughout life [7,8]. Therefore, many authors recommend resection, regardless of the presence or absence of symptoms [8–10]. In contrast, EPS is often asymptomatic and is less likely to become infected later in life. Some authors advocate observation for asymptomatic cases or recommend intervention only after
symptoms appear [11–13]. Even though the surgical results are satisfactory and the complication rates are generally low, pneumothorax, hemothorax, and emphysema are recognized problems. Moreover, the possibility of infection, destruction of the normal pulmonary parenchyma, and malignant transformation have been reported by other groups [4,10,14].

Pulmonary sequestration has previously been reported most commonly in association with CCAM/CPAM type 2, and there has been a misconception that PPB is a preexisting CCAM/CPAM that has undergone malignant transformation [15,16]. Approximately 50% of EPSs contain multiple small cysts, measuring up to 2.0 cm, similar to CPAM type 2, and these are generally termed “hybrid lesions.” However, recent studies by the International Pleuropulmonary Blastoma Registry have indicated that CPAM type 2 does not undergo malignant transformation. In fact, these cystic lesions are assumed to be secondarily developed lesions associated with in utero airway obstruction and/or atresia, a pathological entity quite distinct from CPAM type 2 [15,17]. Although the risk of malignant transformation of the so-called “hybrid lesions” has been rejected, the occurrence of PPB associated with extralobar lung tissue, likely commonly EPS, as in our patient, cannot be ignored.

The development of minimally invasive surgery has increased guardians’ willingness to consent to surgical resection in these children because scarring is minimized, there is less pain, and the hospital stay is shorter than for thoracotomy [15]. Transcatheter arterial embolization (TAE) has recently been performed on selected patients with pulmonary sequestration. Some TAE studies have shown good results, with complete regression of the lesion, although the long-term efficacy and safety of the treatment have not yet been established [18]. However, guardians are often reluctant to consent to surgery, especially when the patient is asymptomatic. In these cases, TAE may be a treatment option, although clinicians should note that no tissue is available for pathological examination after TAE. When considering the treatment options for these lesions, although they are extremely rare, the possibility of PPB associated with extralobar lung tissue (possibly EPS) should be considered.

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References