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Pleuropulmonary blastoma in an adolescent

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Abstract

Primary pulmonary malignancies are rare in childhood. The most common, pleuropulmonary blastoma (PPB), has an incidence of 25–50 cases per year in the United States (Knight and et al., 2019) [1]. The majority of children are diagnosed with PPB before the age of four years. PPB is divided into subtypes I, Ir (type I-regressed), II, and III, which correlates to the age of diagnosis and patient prognosis [2,3]. Here we report an unusual presentation of PPB in a teen-aged female who presented with a one month history of a non-productive cough.

Keywords

Cancer; Pleuropulmonary blastoma; Pediatric

1. Case report

A 13-year-old female with a past medical history significant for asthma presented to the emergency department for evaluation of a non-productive cough of one month duration and an abnormal chest radiograph showing left lung hyper-expansion with mediastinal shift, remodeling of the sternum, and no acute pneumonia. The child had been previously evaluated several times for cough symptoms and treated with anti-allergy medications, cough suppressants and a course of antibiotics. Family history was unremarkable. Physical examination was significant for absence of breath sounds on the left.

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Authorship

All authors at test that they meet the current ICMJE criteria for Authorship.

Patient consent

Consent to publish the case report was not obtained. Our institution is except from the requirement of an IRB for a case report.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Routine laboratory investigations were normal. Chest CT scan demonstrated a large, gas-filled, septated lesion occupying the majority of the left lung with pronounced rightward mediastinal shift. The lesion demonstrated an internal nodular, lobulated solid focus which raised the concern for a potential underlying neoplasm, rather than a congenital pulmonary airway malformation or CPAM (Fig. 1).

The child was admitted to the hospital and underwent a video-assisted thoracoscopic resection of the left lower lobe lung lesion. Gross examination showed a 14.0 × 11.5 × 3.7 cm primarily cystic lesion with an irregular granular to nodular mass-like area of white hard tissue measuring 3.5 × 2.0 × 1.8 cm (Fig. 2).

Microscopically, the mass was noted to be a multi-cystic lesion located in the periphery of the lung with numerous well differentiated chondroid/cartilaginous nodules present in many septa. There was no pleomorphism, atypia, or mitotic activity (Fig. 3). Histopathologic diagnosis was noted to be pleuropulmonary blastoma, type Ir.

The patient had an uncomplicated postoperative course. Oncology consultation was obtained, and due to pathology indicating type Ir, chemotherapy was not administered to the patient. She is being closely observed for disease recurrence with serial imaging and is well at 9 months follow-up.

2. Discussion

Lung malignancies in children are exceedingly rare, representing 0.5–1% of primary lung cancer. PPB is the most common pediatric primary pulmonary neoplasm, usually arising from either the lung or pleura. The majority of patients with PPB present before 4 years of age [2]. The patient in this report was rare, presenting with PPB as an adolescent. There have been a few other cases reports in adolescents and young adults [4] and the disease has been reported in adults up to 40–50 years old in a very few cases [5,6].

PPB is divided into subtypes I, Ir, II, and III. Type I is a purely cystic lesion, and type Ir is a purely cystic tumor which does not have a primitive cell component [3]. Type IR makes up 23% of the total type I patient population and type Ir patients are diagnosed at a later age; type I patients median age is 8 months whereas the type Ir patients median age is 46.5 months [1]. Type II tumors have cystic and solid components, and type III are purely solid lesions [3]. Type I/Ir lesions account for 15–20% of PPB while type II and III account for 80–85% [7]. Type I and Ir tumors carry a good prognosis with an 80–90% 5-year disease-free survival. All reported deaths associated with type I occurred with progression to type II or III and the incidence of progression has been reported to be approximately 10%. Types II and III tumors have worse 5-year disease free survival at less than 50% [2].

PPB’s initial stage is a cystic lesion, which can make differentiating PPB from congenital pulmonary airway malformation difficult [8]. Additional differential diagnoses include synovial sarcoma and fetal lung interstitial tumor [2]. PPB is associated with the germline mutation in DICER1, a member of the ribonuclease III family [1]. The patient in this report was negative for the DICER mutation.

Presenting symptoms depend on the type of lesion, ranging from fever, respiratory distress, chest pain, and pneumothorax [2]. Imaging may have varied findings, including a single cyst, multi-cystic lesion, or solid lesions with or without an associated cystic component [7]. Approximately 9% of type II and III lesions present with metastasis, which are much less common in type I tumors. The most common sites of metastasis are the brain and bone and less commonly the liver. Because of the increased risk of metastatic disease in patients with types II and III, recommended evaluation for these patients includes an MRI of the brain and a bone scan [2].

Treatment of PPB depends on the tumor type. Patients with type I tumors undergo surgical resection for diagnosis and treatment. The use of chemotherapy in patients with types I or Ir has not been shown to decrease the rate of progression to types II or III or impact overall survival [2]. For patients with types II or III, systemic chemotherapy is recommended, and the recommended regimen includes Ifosfamide, Vincristine, Actinomycin-D and Doxorubicin [9]. Surgery is an essential component of treatment for these patients as well, and multiple thoracotomies may be required to achieve complete resection. Radiation has been used for patients with unresectable residual primary tumors, though it has not been shown to affect survival [2].

Our patient presented with a cough and an abnormal chest radiograph. She was diagnosed with type Ir PPB, which is less common than type II and III, but carries a better prognosis. Patients with symptomatic type I PPB typically present with respiratory distress with or without an associated pneumothorax [2], which our patient did not demonstrate. Her age, 13 years old, is also an unusual time of presentation for PPB. This case demonstrates that pediatric providers must consider PPB in the differential diagnosis, even in older children, when an abnormal chest CT scan is encountered.

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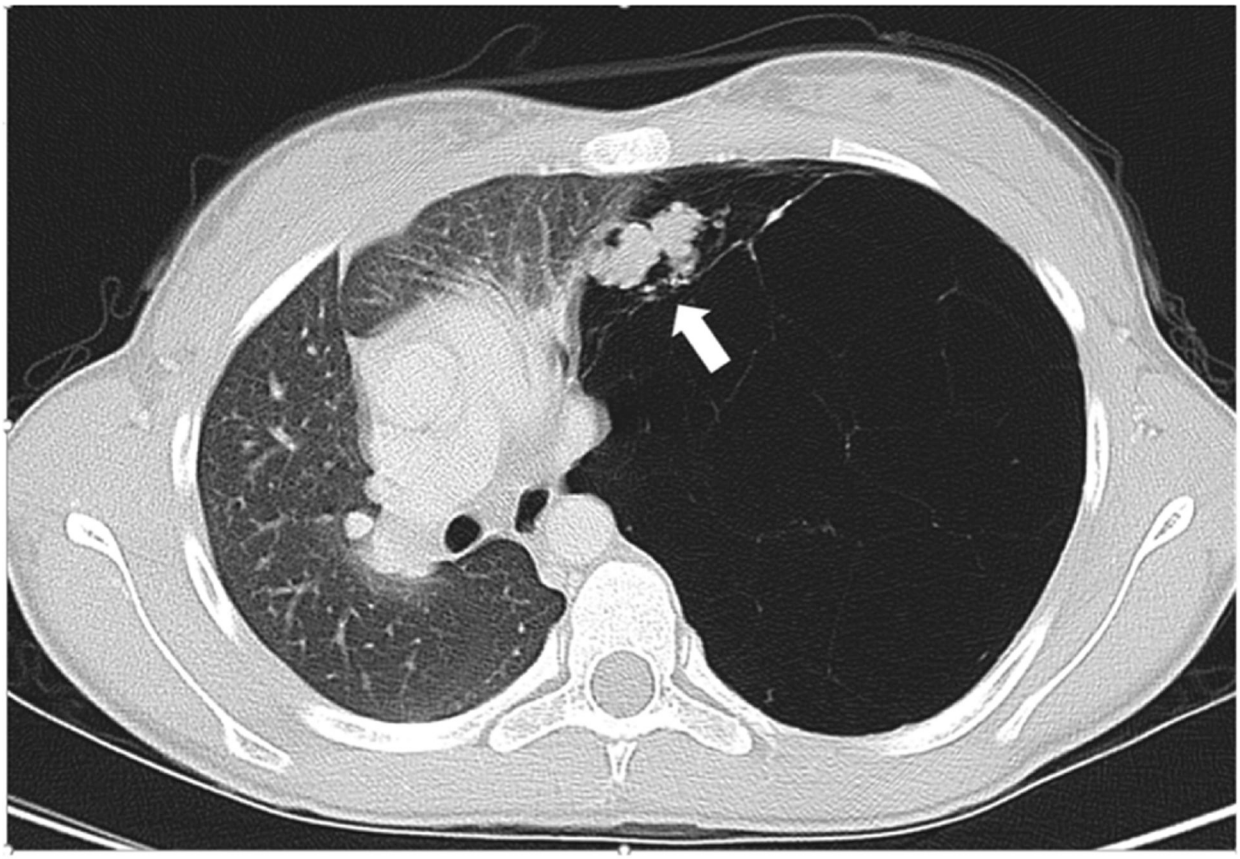

Fig. 1.

Axial CT of the chest lung window Large cystic lesion with internal thin septations exerting marked mass effect and secondary rightward mediastinal shift An internal single lobulated solid nodule (white arrow) raised the concern for a potential underlying neoplasm.

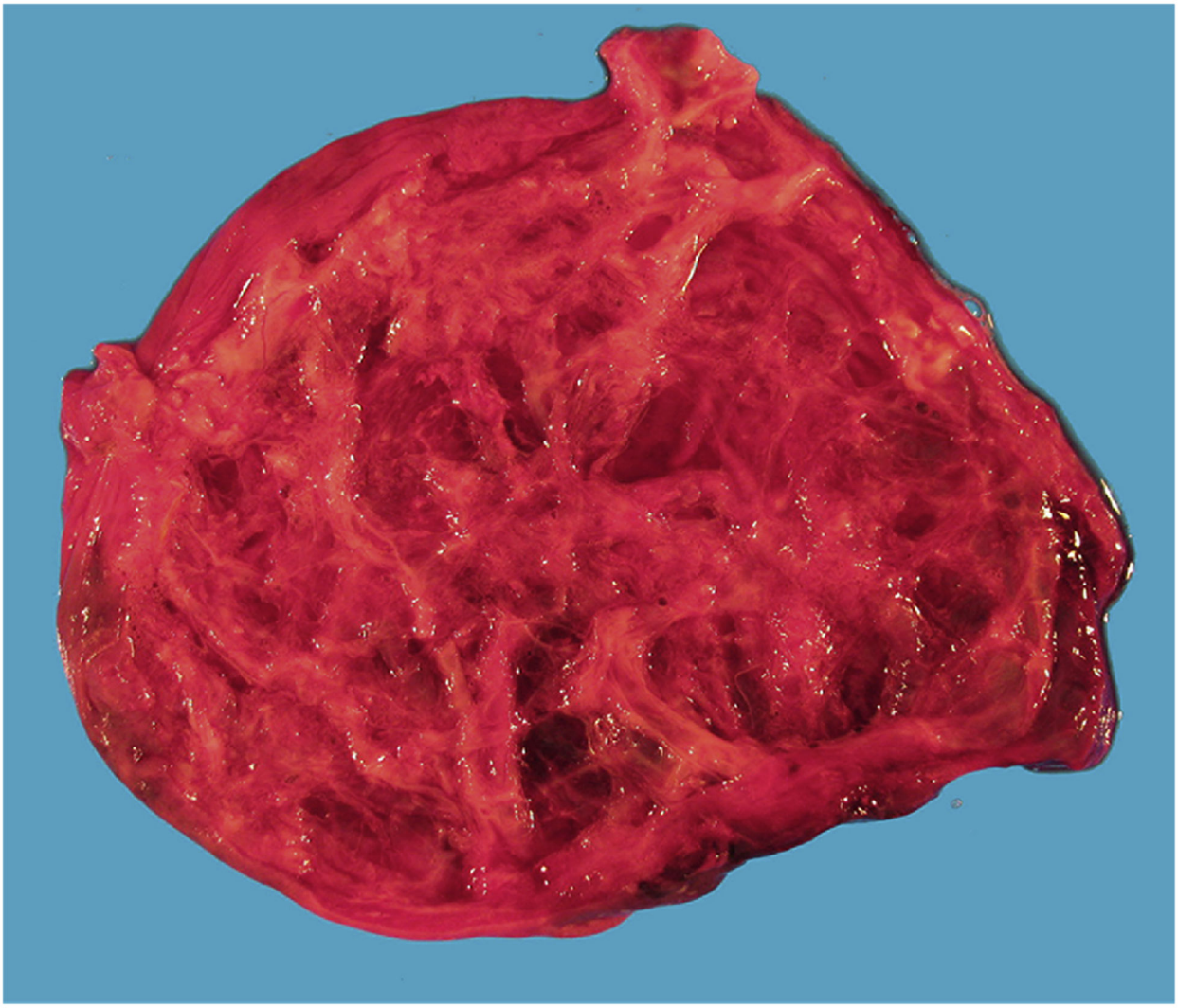

Fig. 2.

The cut surface of this 121 g left lower lobe lung speciman shows an 11.5 cm mulitilocular cystic lesion with small coarse and fine granular chondroid nodules in the walls.

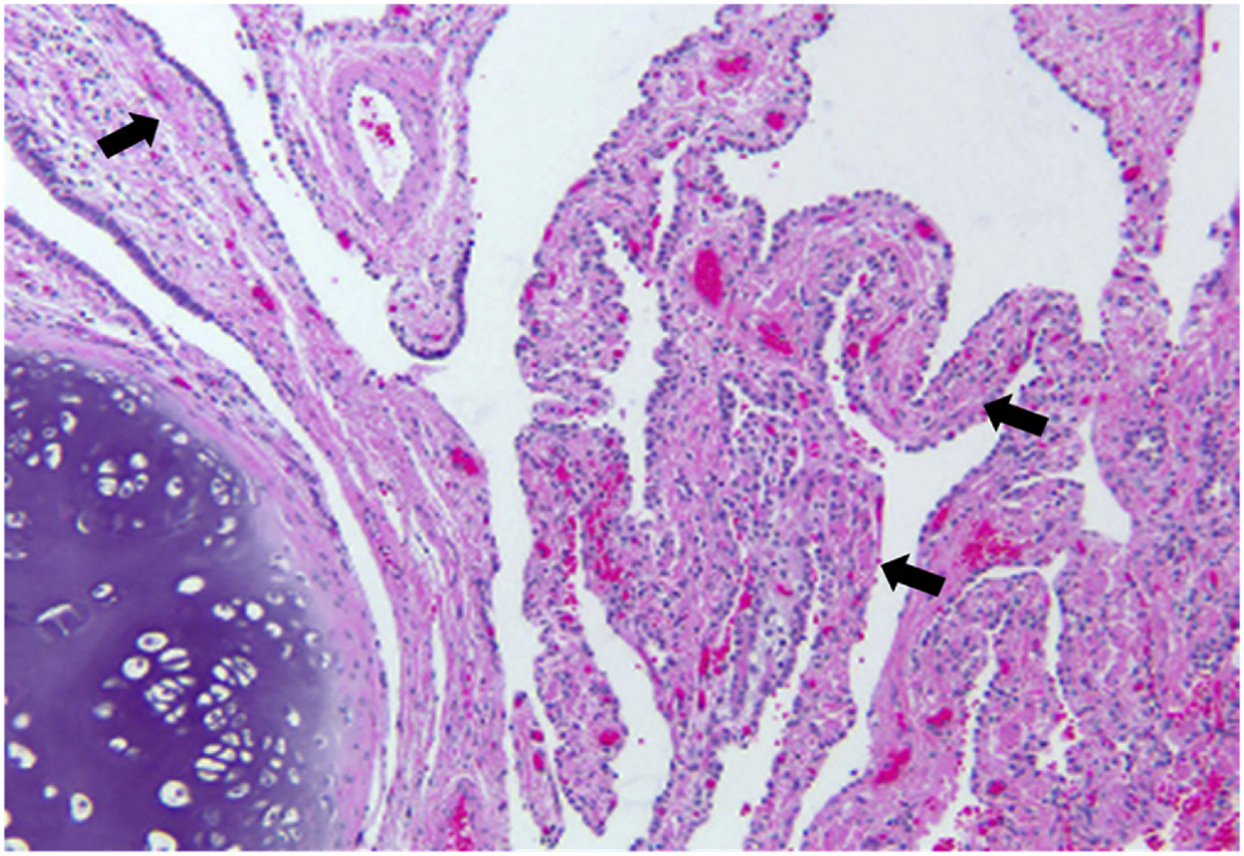

Fig. 3.

The thin-walled cysts contained scattered well-differentiated chondroid nodules, slender bands and bundles of smooth muscle (black arrows) (without a neoplastic rhabdomyosarcomatous cellular infiltrate) and were lined by respiratory epithelium with variably prominent type Ⅱ pneumocytes.