

Letter to the Editor

## Pleuropulmonary blastoma: report of a case presenting with spontaneous pneumothorax

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A 2<sup>1</sup>/<sub>2</sub>-year-old boy was referred to the Department of Thoracic Surgery with pneumothorax. He had a history of difficulty in breathing, cough and fever for the last 6 months. Prior to admission, he had undergone antibiotic treatment for pneumonia, which was diagnosed on chest radiogram. He was dyspneic and had leukocytosis. Chest radiograms showed right tension pneumothorax and a cystic mass lesion in the right lower lobe, the radiologic appearance simulating an air crescent sign. A chest tube was inserted into the right hemithorax, resulting in lung expansion. Thorax CT confirmed a large solid mass with necrotic areas, filling the right inferoposterior hemithorax. Magnetic resonance examination revealed a supradaphragmatic, suggestively extraparenchymal, mass compressing the lower lobe. Bronchoscopy was not performed since the tumor was peripherally located. At thoracotomy, a predominantly solid and partially necrotic mass with extraparenchymal extension was found to originate from the visceral pleura, extending posteriorly to the mediastinum and inferiorly to the diaphragm. The mass involved the diaphragmatic surface of the posterobasal segment through a narrow base. Although firm adhesions were present, involvement of the parietal pleura was not observed. The mass was removed completely with local excision and wedge resection. Lobectomy was not required. The resected 7 × 7 × 3 cm mass was histopathologically evaluated to present with morphological characteristics of pleuropulmonary blastoma (PPB). The patient did well in the postoperative period and was discharged on the 7th postoperative day. Eight courses of cisplatin (100 mg/m<sup>2</sup>) and etoposide (100 mg/m<sup>2</sup>) were administered postoperatively at an oncology center. He is well and free of recurrence at 10 months following surgery.

PPB is defined as a distinctive pulmonary and/or pleural tumor of childhood with blastomatous and sarcomatous features without any epithelial component, differentiating it from the classical adult type pulmonary blastoma [1]. It is pathologically divided into cystic, solid and mixed types [2]. PPB is considered within the category of disembryonic neoplasms such as Wilms' tumor, neuroblastoma and hepatoblastoma [1]. The etiology and predisposing factors are unclear. However, PPB arising in the presence of cystic pulmonary disease have been reported [1]. In the present case, there was no cystic remnant within the tumor and radiological cystic appearance was principally due to tumor necrosis.

The common presenting symptoms in PPB are respiratory distress, fever, chest pain, cough, anorexia and malaise [3]. PPB may also present with pneumothorax. Pneumothorax was reported in those cases associated with cystic pulmonary disease [4,5]. In the present case, tension pneumothorax might have resulted from spontaneous rupture of the tumor.

The recommended initial treatment for PPB has been complete surgical excision. However, invasion of the surrounding structures and extreme friability of the tumor may prevent complete excision. The prognosis is usually poor. Gender, pathological subtype, tumor size, extent of surgical resection, presence of necrosis and adjuvant treatments such as radiotherapy and chemotherapy have been reported not to influence survival significantly. The best indicators of long-term survival are the presence of parenchymal involvement alone and the absence of pleural or mediastinal involvement [3].

### References

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