Classic Pulmonary Blastoma: A Subtype of Biphasic Pulmonary Blastoma

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We report a rare case of classic pulmonary blastema (CPB) without recurrence for 3 years after the operation. A 70-year-old man presented with cough and sputum for a month. Chest computed tomography (CT) showed a 5cm-sized mass in the right middle lobe. Bronchoscopic examination was performed, and the mass was suspected as adenocarcinoma of the lung. Right middle lobectomy and lymph node dissection were performed. The pathologic histology diagnosis was classic pulmonary blastoma, a subtype of biphasic pulmonary blastoma.

Keywords: classic pulmonary blastoma, biphasic pulmonary blastoma, β-Catenin

Introduction

According to the new World Health Organization classification, biphasic pulmonary blastomas (BPB) are classified as carcinomas with pleomorphic, sarcomatoid, or sarcomatous elements, which are rare tumors accounting for about 0.25%–0.5% of all pulmonary malignancies. The tumor is histologically biphasic with a mixture of immature epithelial and mesenchymal components mimicking well-differentiated fetal adenocarcinoma. Two subtypes have been proposed in cases of BPB.

Case

A 70-year-old male presented with cough and sputum for a month. Chest x-ray revealed a large mass in his right lower lung field. His chest computed tomography (CT) showed a uniformly enhanced tumor about 7cm in diameter in the right S4 (Fig. 1). Bronchoscopic examination was performed, and the mass was suspected as adenocarcinoma of the lung. Right middle lobectomy and lymph node dissection were performed. Grossly, the tumor was relatively well-defined, 7.0 × 5.5 × 4.2 cm in size, and light-yellowish on cut-section; there was no evidence of hemorrhage or necrosis (Fig. 2). Histologically, the tumor was composed of multiple ductal structures and stromal components comprising spindle cells. Morula formation was noted within the ducts. The interstitium was primarily composed of pleomorphic or short spindle cells. Furthermore, prominent nuclear atypicality and abnormal mitosis were noted, but no mesenchymal components suggestive of rhabdomyosarcoma or chondrosarcoma were identified (Fig. 3a). Immunohistochemical inspections revealed positive for β-catenin, thyroid transcription factor-1 (TTF-1), MIB-1, epithelial membrane antigen (EMA), Cytokeratin (AE1/AE3), VIMENTIN (Fig. 3b–3e).

The pathologic histology diagnosis was Classic pulmonary blastoma, a subtype of biphasic pulmonary blastoma. Passage was excellent after the operation. He left hospital on the 8th day after the operation. Although postoperative adjuvant chemotherapy was not undertaken at his request, he was good without recurrence for 3 years.
Fig. 1  Chest computed tomography scan showed a uniformly enhanced tumor, of about 7 cm in diameter, in the right S4.

Fig. 2  The tumor cut-surface showed relatively well-defined, 7.0 × 5.5 × 4.2 cm in size, and light-yellowish on cut-section; there was no evidence of hemorrhage or necrosis.

Fig. 3  

a: the tumor was composed of multiple ductal structures and stromal components comprising spindle cells. Morula formation was noted within the ducts. (HE × 4)
b: β-Catenin is expressed in both epithelial and mesenchymal components of the tumor.
c: TTF-1 is expressed in epithelial components of the tumor.
d: Cytokeratin (AE1/AE3) is strongly expressed in epithelial components of the tumor.
e: VIMENTIN is expressed in mesenchymal components of the tumor.

TTF-1: thyroid transcription factor-1
Discussion

The pathogenesis and clinical pathology of BPB remained unknown for a long time. However, tumors conventionally diagnosed as BPB primarily during adulthood have been proposed to be composed of two different tumors: [1] classic pulmonary blastoma (CPB), in which fibroblast-like mesenchymal cells are produced from low-grade fetal lung adenocarcinoma (L-FLAC) and well-differentiated fetal adenocarcinoma (WDFA) through an epithelial-mesenchymal transition (EMT); and [2] granulomatous subtype of carcinosarcoma, in which sarcoma components are produced from high-grade fetal lung adenocarcinoma (H-FLAC) and clear cell adenocarcinoma (CCA) with fetal lung features through EMT. CPB is most frequent in patients in their 30s and 40s, while the granulomatous subtype of carcinosarcoma mainly affects elderly male patients.

In the present patient, the tumor was difficult to differentiate from the granulomatous subtype of carcinosarcoma because of the prominent nuclear atypicality. However, the tumor was diagnosed as CPB based on the morula formation, the β-catenin staining showing positive epithelial and mesenchymal components, and prominent nuclear-cytoplasmic pattern particularly in the morula-like cells. The epithelial components were TTF-1 positive, thereby ruling out granulomatous subtype of carcinosarcoma.

Preoperative diagnosis of pulmonary blastoma is difficult because the expansive tumor growth in the bronchial tube interferes with transbronchial diagnosis and the epithelial and mesenchymal cells cannot be distinguished by biopsy. Cytology may only determine the malignancy in some patients but usually identifies adenocarcinoma. In the present patient, exfoliative cytodiagnosis raised a suspicion of adenocarcinoma, but the subsequent examination of the resected specimen showed stroma-like cells with a spindle-shaped nucleus.

Surgery is the first treatment of choice for CPB, but the prognosis is poor. Two-thirds of CPB patients die within 2 years after the diagnosis. The 5-year survival rate is 16%. Our patient is alive and well without recurrence or metastasis at 3 years postoperation.

Conclusion

CPB is a rare tumor, and many of its features remain unsolved. Thus, clinical data of CPB patients need to be accumulated. Since no effective treatment other than surgery or standard treatment is currently available, treatment should be carefully considered according to individual patients.

Disclosure Statement

I declare I have no conflict of interest in connection with this paper.

References