Clinical Case Report

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Clinical and pathology analysis of 1 case of adult pleural pulmonary blastoma

A case report

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Abstract

Rationale: Pulmonary blastoma is a rare primary lung cancer that can be categorized into adult type and child type. The clinical symptoms and imaging features of pulmonary blastoma are nonspecific, making it difficult to diagnose preoperatively. Postoperative pathology with immunohistochemical staining can help diagnosis.

Patient concerns: A 53-year-old male had chest tightness and shortness of breath.

Diagnoses: The patient was diagnosed as pleural pulmonary blastoma based on computed tomography (CT) scan, pathology, immunohistochemistry, and molecular pathology. CT examination showed solid mass on the upper lobe of the left lung Intraoperative observation found that tumor tissue was gray with tough texture. The surrounding lung tissue showed AE1/AE3 (+), Vimentin (+), and CD34 (+) staining. No epidermal growth factor receptor gene mutation was detected.

Interventions: The left lobe resection plus mediastinal lymph node dissection were performed. After the operation, patient received paclitaxel combined with nedaplatin chemotherapy for 4 times.

Outcomes: Four months later, left pleural metastasis, and mediastinal lymph node metastasis was found. The patient died 15 months later.

Lessons: Pleural pulmonary blastoma is a malignant tumor with rare pathological features that is easy to relapse and metastasis with poor prognosis. Surgical treatment preferably, lobectomy plus mediastinal lymph node dissection, is the first treatment option. The overall prognosis is poor.

Abbreviations: CEA = carcinoembryonic antigen, cgA = chromogranin A, CK = cytokeratin, EMA = epithelial membrane antigen, NSE = neurogenic specific enolase, PPB = pleural pulmonary blastoma, SMA = smooth muscle actin.

Keywords: clinical, pathology, pleural pulmonary blastoma

1. Introduction

Pleural pulmonary blastoma (PPB) is a rare malignant tumor of the lung. It consists of mesenchymal and/or epithelial components, which is similar to the structure of the embryonic lungs and accounts for about 0.25 to 0.5% of all primary lung malignancies.[1] Pulmonary blastoma has nonspecific clinical symptoms and imaging features and is difficult to be diagnosed preoperatively. Depending on the onset age, PPB can be divided into 2 types: adult and child type.[2] Adult type can be histologically categorized as biphasic pulmonary blastoma and monophasic pulmonary blastoma. The former contains the original malignant epithelial cells and the original mesenchymal tissues. The latter, which is also called epithelial or well-differentiated adenocarcinol, contains only malignant epithelial components. Pediatric pleuropulmonary blastoma is a monophasic pulmonary blastoma, containing only the original mesenchymal tissue. In this study, we analyzed the clinical characteristics of 1 adult PPB, and discussed its histopathological characteristics.

2. Case presentation

Prior written and informed consent were obtained from this patient and the study was approved by the ethics review board of Xinjiang Medical University Affiliated Cancer Hospital.

A 53-year-old male was admitted because of chest tightness and shortness of breath for 2 months. Since the flu 2 months earlier, patient had gradually increasing chest tightness, chest pain, and cough, but without sputum, hemoptysis, fever, or night sweats. Chest CT scan showed huge lump shadow on left lobe with the size of 14.5 × 10.3 cm. The boundary was clear with uneven density. Flaky shadows with slightly high density were observed. The CT value of plain scan was about 27 HU. Under enhanced scan, the CT value of the arterial phase was about 33 HU and that of the vein phase was about 40 HU (Fig. 1). The patient had good heart and lung function, and no tumor distant metastasis sign was detected. Patient had left upper

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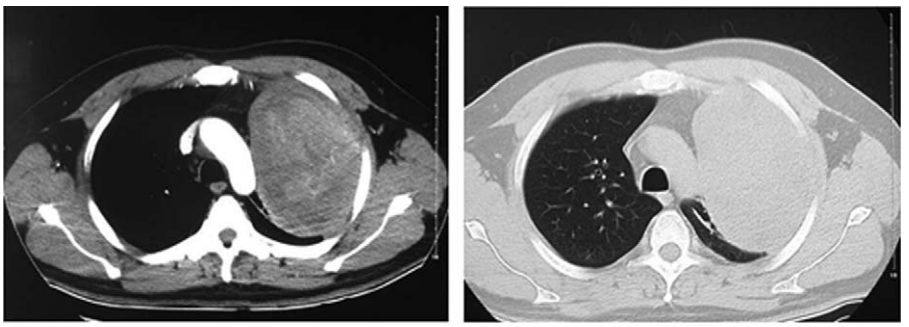

Figure 1. (A) Vertical diaphragm window showed on the upper left lobe of left lung, a mass with clear boundary, uneven density and enhanced after enhanced scan. (B) Lung window showed tumor edge without burring.

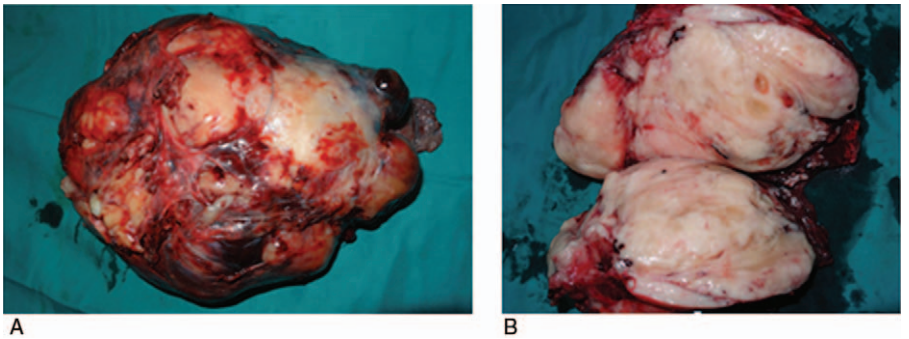

Figure 2. (A) Solid mass in the lung tissue showed clear boundary without capsule. (B) Tumor section was gray with cystic structure.

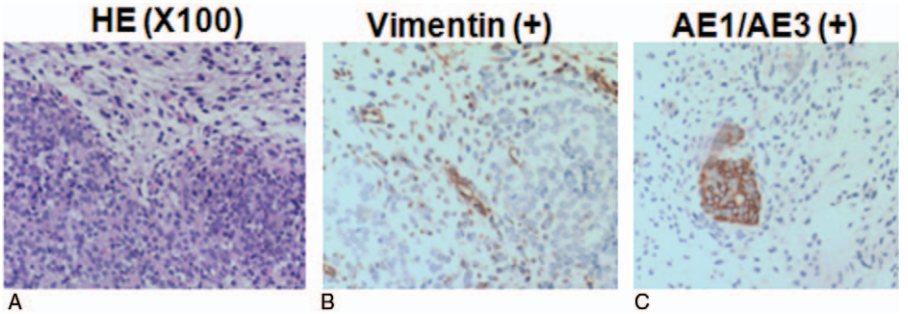

Figure 3. (A) HE staining showed mixed epithelial cells and mesenchymal cells (HE ×100). (B) Positive vimentin immunohistochemical staining (magnification ×100), and (C) positive AE1/AE3 immunohistochemical staining (magnification ×100). HE=hematoxylin-eosin.

lobectomy + mediastinal lymph node dissection under general anesthesia. Postoperative pathology diagnosis showed PPB and no lymph node metastasis was found. The postoperative staging was PT3N0M0 (IIB). After surgery, general examination showed a solid tumor on the upper lobe of the left lung. The tumor was gray and tough. The tumor tissue had clear boundary and the tumor size was 20.0 cm × 8.0 cm × 1.5 cm (Fig. 2). Postoperative immunohistochemical examination showed epithelial cell AE1/ AE3 (+), mesenchymal cell vimentin (+), and vascular CD34 (+), and epithelial cells and mesenchymal cells were in mixed arrangement (Fig. 3). High-resolution melting analysis method was used to detect gene mutations and showed no mutation of epidermal growth factor receptor (EGFR) 18, 19, 20, 21 exons.

After surgery, the patient received 4 courses of paclitaxel + nedaplatin. Four months after surgery, CT scan showed multiple nodules on left pleura, anterior mediastinum, and left anterior intercostal, suggesting metastatic tumor. Therefore, radiotherapy of 42 Gy/21 f was performed. One month later, CT scan showed pleural metastases were still in progression. The patient received further chemotherapy (pemetrexed + cisplatin + bevacizumab) 1 year after the surgery and died half a year later due to multiple organ failure.

3. Discussion and conclusion

PPB is a rare malignancy associated with dysplasia in children.[3] The origin of the tissue is not yet clear, and may be derived from

the lung cancer stem cells.[4] Due to its rarity, there is no report on the incidence of PPB. The International Pneumoblastoma Agency showed about 300 cases reported so far.[5] PPB in adults is more rare. Clinical characteristics of PPB are often nonspecific, presenting as cough, sputum, hemoptysis, chest tightness, shortness of breath, chest pain, and discomfort.[6] These clinical features are associated with tumor size and invasion.[7] In this study, this patient showed chest tightness and shortness of breath. CT scan showed isolated peripheral lesions with clear boundaries, large size, and uneven density.

The pathological presentations of PPB can be divided into simple cystic, cystic, and solid. Under light microscope, they were divided into polycystic type (type I, with capsule wall containing mature respiratory epithelial cells and small and round primary mesenchymal cells under the epithelium), polycystic solid type (type II, showing solid region on the basis of type I and the solid region was composed of naive round or spindle cells and sarcomatoid components), and solid type (type III, composed of naive mesenchymal cells without cystic structure).[8] Immunohistochemistry showed positive staining of cytokeratin (CK), carcinoembryonic antigen (CEA), and epithelial membrane antigen (EMA) in the epithelium[9] and positive staining of Vimentin, Desmin and smooth muscle actin (SMA) in the mesenchyme.[10,11] In some cases, malignant epithelium has primitive endocrine function, and the neurogenic specific enolase (NSE), chromogranin A (cg A), and synaptophysin are positively stained.[12] In this report, there was bi-directional differentiation in PPB, with epithelial-like cells and mesenchymal cells mixing together. Immunohistochemistry showed that epithelial cell AE1/AE3 (+), mesenchymal cell vimentin (+), and vascular CD34 (+). The tumor morphology and immunohistochemical results supported the diagnosis of PPB.

PPB treatment mainly includes surgery, radiotherapy, and chemotherapy. PPB can be highly invasive, so conservative surgery is not recommended. The lobectomy + lymph node dissection is used as routine operation for PPB.[13] Wedge resection should be used in caution.[14] Lymph node metastasis and primary tumor size are associated with poor prognosis.[15] For local progress (IIIB) or advanced (IV) PPB, surgical resections are difficulty to perform.[16] Thus, neoadjuvant chemotherapy should be used first to reduce the tumor stage and then surgical resection should be conducted.[17] However, because of the rarity, definitive treatment is unclear. At present, tumor molecular targeted therapy and individualized medical care are advancing.[18] EGFR gene mutations are closely related to the efficacy of anti-EGFR monoclonal antibody treatment.[19] EGFR gene mutation can be used as a prognostic predictor to guide drug use. In the present study, there was no mutation in EGFR 18, 19, 20, or 21 exons, suggesting that the patient was not sensitive to epidermal growth factor inhibitor drugs.[20] It is reported that PPB may have recurrence or metastasis in a short term, and the 5-year survival rate of most cases are only 45%.[21] This case in this report received chemotherapy and postoperative radiotherapy for 4 courses after surgery, had pleural metastasis 5 months after surgery, and survived for 15 months. However, whether PPB patients can benefit from postoperative radiotherapy and whether postoperative adjuvant chemotherapy can reduce distant metastasis are unknown. Further study with large sample size is needed.

In conclusion, the prognosis of PPB is poor and is associated with its pathological type, tumor stage, and tumor site. Surgical treatment is the first option. Because of its high invasiveness, lobectomy plus lymph node dissection is preferred. For different staging and types of pulmonary myoblastoma, whether postoperative radiotherapy can improve local control rate and postoperative chemotherapy can reduce distant metastasis requires large-scale randomized controlled trial.

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