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Case report

Radical resection of a pulmonary blastoma involving the pulmonary artery

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Abstract

Pulmonary blastoma is a rare malignant lesion with a poor prognosis. We described a case of a 47-year-old woman with a large biphasic pulmonary blastoma, involving the left pulmonary artery. Under cardiopulmonary bypass, it was treated with radical left intrapericardial pneumonectomy and pulmonary thromboendarterectomy. Subsequent chemotherapy and radiotherapy was used. Three years postoperatively, the patient was clinically and radiologically free of tumor.

Keywords: Lung cancer surgery; Cardiopulmonary bypass

1. Case report

Pulmonary blastoma is a rare malignant neoplasm of the lung comprised of epithelial and/or mesenchymal elements [1]. The tumor was originally described by Barnard and Spencer [2,3]. Pulmonary blastoma is a malignant lesion with a poor prognosis. Therapy should include surgical resection and subsequent chemotherapy and radiotherapy. We reported a 3-year complete remission produced by radical surgical resection followed by adjunctive therapy in a pulmonary blastoma involving pulmonary artery.

A 47-year-old woman presented with dry cough that increased over 1 month. There was no history of chest pain, fever or weight loss. Physical examination was unremarkable. The patient denied a history of smoking. Family history was free of cancer. The X-ray and computer tomographic scan of chest showed a large mass occupying 1/3 of left hemithorax, which had sharp margins without surrounding inflammation (Fig. 1). Bronchoscopy revealed the narrowed upper and lower bronchi, and no evidence of an intrabronchial lesion. Percutaneous and transbronchial biopsies did not reveal the diagnosis. Distant metastases were excluded by abdominal ultrasound scan, brain magnetic resonance imaging and bone emission computed tomography.

A cervical mediastinoscopy was performed to evaluate the right, left and subcarinal paratracheal lymph nodes. The results of anatomopathologic analysis performed by freezing were negative. Then the patient received left posterior lateral thoracotomy, entering the thoracic cavity through the fifth intercostal space. The inventory of the thoracic cavity showed no significant adherence between the upper lobe and the parietal pleura. A compact irregular mass of 15 cm × 12 cm × 9 cm was localized in the upper left pulmonary lobe. There were no evidences of extension of this mass to the chest wall or pericardium and no implants on the pleura. There were no obviously enlarged mediastinal nodes. But the inventory of left lung hilum showed that the tumor had invaded the left pulmonary artery. After pericardiotomy, a tumor thrombus was found in the left pulmonary artery which exceeded the bifurcation of pulmonary trunk, and extended to right pulmonary artery for 1 cm. Therefore, a cardiopulmonary bypass was performed using the femoral artery-femoral vein approach. Under cardiopulmonary bypass, pulmonary thromboendarterectomy and arterioplasty were sequentially performed, after blockade pulmonary trunk and proximal part of right pulmonary artery. In brief, we made an incision on the wall of pulmonary artery, and the thrombus was removed completely. Then the PA defect was closed by continuous sutures of the left wall of the bifurcation with 5-0 prolene sutures. The mass was removed en bloc through radical left intrapericardial pneumonectomy.

Histology confirmed pulmonary blastoma of biphasic type (Fig. 2), bronchial stump and lymph node free of neoplasia. The length of the thrombus was about 5 cm diagnosed as neoplasm metastatic by anatomopathological exam. The pathological stage was T4N0M0. Immunohistochemistry was positive for vimentin and actin in the mesenchymal elements

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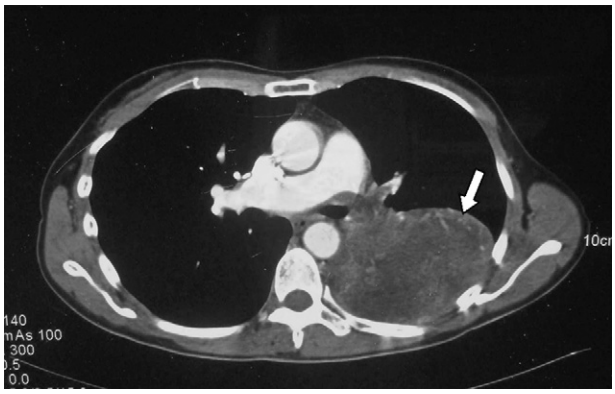

Fig. 1. Computed tomographic scan of the chest show the mass in the left upper lobe (arrow).

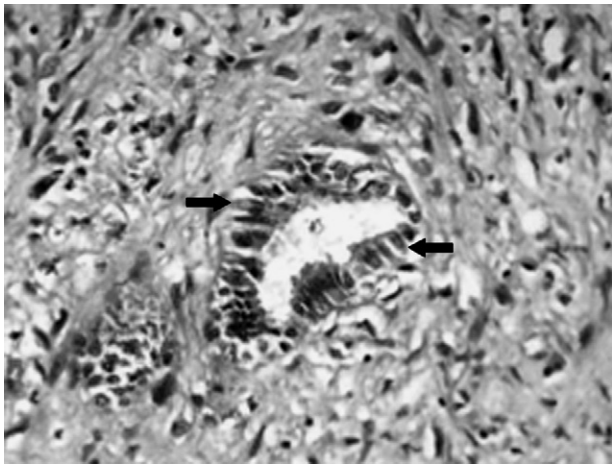

Fig. 2. Biphasic histology of malignant glandular elements (arrow) surrounded by embryonic mesenchyme (H&E, original magnification, ×200).

of the tumor. S-100 and EMA were positive in the epithelial elements.

The patient discharged from hospital following an uneventful recovery on postoperative day 12. Four weeks postoperatively, the patient was treated with three cycles of a systemic chemotherapy applying carboplatin, etoposide and isofosfamide daily for 3 consecutive days at 3-week intervals. Three months postoperatively, the patient received radiotherapy to a total of 6000 Gy in 30 fractions over 8 weeks, covering the areas of the tumor bed, left hilum, mediastinum, and supraclavicular areas.

After the termination of her treatment, the patient was followed up every 6 months. At last follow-up, 3 years postoperatively, the patient was clinically and radiologically free of tumor.

2. Comment

The etiology and predisposing factor are not still fully understood. In the literature, there is a strong correlation between smoking and pulmonary blastoma [4,5]. The tumor may occur at all ages but predominantly common in adults, with the peak incidence in the fourth decade of life [4]. Compared to non-small cell lung cancer, the patients may be asymptomatic at presentation. Preoperatively, pathologic confirmations with noninvasive procedures are of low yield [6]. Therefore, they were usually diagnosed in advanced stages.

There are two histological types of pulmonary blastoma: monophasic and biphasic pulmonary blastoma [4]. Francis reported an overall 5-year survival of 16% [7]. Poor prognostic features for pulmonary blastoma include biphasic histology, the size of mass (greater than 5 cm), and metastasis [4]. Our patient's tumor was biphasic and had another poor prognostic factor, a size >5 cm in dimension. They were all dire prognostic signs.

The primary treatment of pulmonary blastoma is radical surgery. Surgical resection of lung cancer involving great vessels is uncommonly performed because of the potential morbidity and mortality for an unknown probability of significant palliation or cure. At the exploratory thoracotomy, we found the involvement of left pulmonary artery and the pulmonary artery tumor thrombus. From the experience of this case, we thought that the tumor invading the pulmonary artery could be safely resected under cardiopulmonary bypass, which resulted in significant palliation and prolonged survival.

Because of lack of data, adjunctive therapy for pulmonary blastoma is still controversial. Our patient with the tumor greater than 5 cm and biphasic histology, but also with invasion of the pulmonary artery, was believed to be at high risk of recurrence. Therefore, it may be reasonable to treat her in combination with chemotherapy and radiotherapy after operation. Because of no standard or known effective regimen for pulmonary blastoma, the selection of chemotherapeutic agents was empiric in this patient.

To our knowledge, this study reported the first biphasic pulmonary blastoma involving the pulmonary artery that had mid-term remission after radical surgical resection followed by adjuvant therapy. For this patient, the multimodality therapy might be effective.

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