

A True Pulmonary Carcinosarcoma that Required Diagnostic Differentiation from a Pleomorphic Adenoma: A Case Report

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Pulmonary carcinosarcoma is a rarely encountered tumor. We treated a patient who had an intrabronchial polypoid lesion that required a diagnostic differentiation from epithelial-mesenchymal mixed neoplasms inclusive of pleomorphic adenoma, and that was diagnosed by immunohistochemical staining to be a true carcinosarcoma. A 69-year-old man underwent left pneumonectomy in November 2000 with a diagnosis of atelectasis resulting from a tumor obstructing the left lower lobar bronchus, and also a lung abscess. The tumor was initially diagnosed as pleomorphic adenoma, since it contained both benign-looking epithelial and mesenchymal elements, but immunohistochemical staining demonstrated myoglobin-positive rhabdomyosarcomatous elements along with cytokeratin-positive squamous cell carcinoma elements. A definite diagnosis of pulmonary carcinosarcoma was confirmed. (Ann Thorac Cardiovasc Surg 2009; 15: 42–45)

Key words: pulmonary carcinosarcoma, pleomorphic adenoma, immunohistochemical staining, lung abscess, pneumonectomy

Introduction

Carcinosarcoma is a rare tumor composed of both malignant epithelial and mesenchymal elements and should be differentiated from several epithelial-mesenchymal mixed tumors of the lung. We describe a case of intrabronchial polypoid tumor that was diagnosed as true carcinosarcoma based on the results from immunohistochemical staining to make a differentiation from other epithelial-mesenchymal mixed tumors, especially pleomorphic adenoma.

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Case

A 69-year-old male demonstrated shortness of breath, generalized malaise, anorexia, and a fever early in October 2000. He was admitted to the hospital for a medical checkup. Hematological data indicated pronounced inflammation. A chest X-ray showed left-sided pleural effusion (Fig. 1). A bronchoscopic examination revealed a white mass on the mucosa at the entrance to the left lower lobar bronchus (Fig. 2), and a microscopic examination of the biopsy specimen showed inflamed tissue with atypical squamous metaplastic cell fragments. Chest computed tomography (CT) showed pulmonary atelectasis in the left lower lobe (Fig. 3). The patient subsequently suffered from a persistent high and spiky fever and underwent an emergency operation in November 2000 with a diagnosis of uncontrollable lung abscess resulting from some type of polypoid tumor. The surgical procedures were determined based on the following findings. First, a chest CT revealed total pulmonary atelectasis in the left lower lobe, and the size of the tumor and its extension could not be determined.

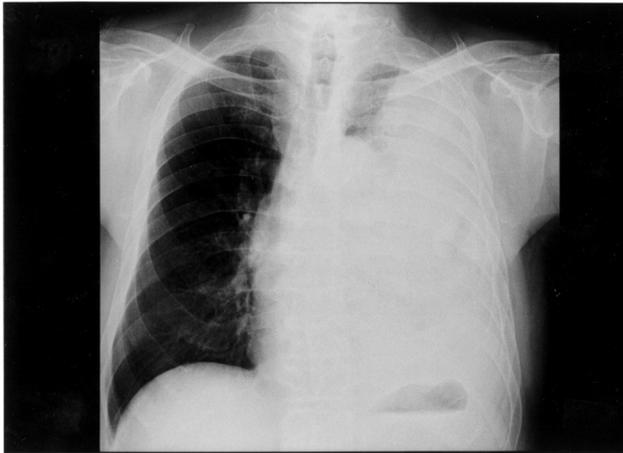


Fig. 1. A chest X-ray showed significant left-sided pleural effusion.

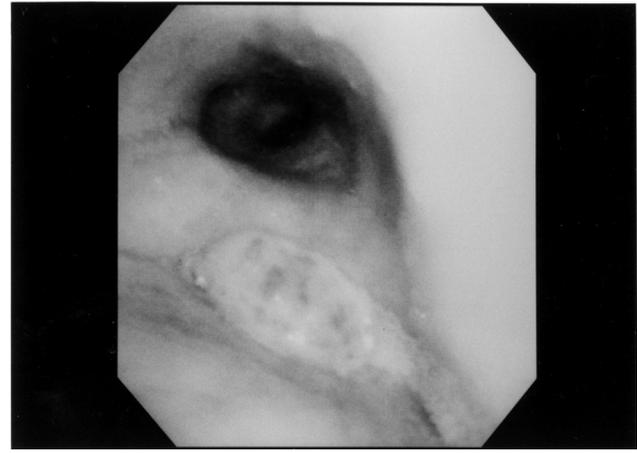


Fig. 2. A bronchoscopic examination revealed a white mass on the mucosa at the entrance to the left lower lobar bronchus.

Second, the bronchoscopic findings suggested that at least a sleeve resection of the lower lobe would be necessary, though the tumor was possibly benign. Third, the chest X-ray results and a marked fever indicated that inflammation was present across the left thoracic cavity with possible significant adhesion. Lastly, the patient demonstrated a decreased glucose tolerance, which led to poor glucose control prior to surgery. These findings indicated that a left total pneumonectomy would be associated with a smaller risk than a sleeve resection. It was also decided that the total pneumonectomy would include the severing of both the pulmonary artery and the superior pulmonary vein by a midsternal incision, followed by severing of the inferior pulmonary vein by a left posterolateral thoracotomy.

The chest was opened by a midsternal incision, and the main trunk of the left pulmonary artery and the superior pulmonary vein were then severed within the mediastinum, using an automatic suture device. This was followed by severing the inferior pulmonary vein and the primary bronchus through a left posterolateral thoracotomy combined with a thoracotomy at the level of the fifth rib. An additional posterior thoracotomy at the eighth intercostal space was performed to detach the adhesions to the diaphragm to complete the left pneumonectomy. The stumps were thereafter reinforced with intercostal muscle flaps. The tumor was found to have arisen from the left lower bronchial stem, producing an arborescent obstruction of the bronchus, and the left lower lobe was almost entirely filled with empyematous fluid.

The pathological examination revealed several things,

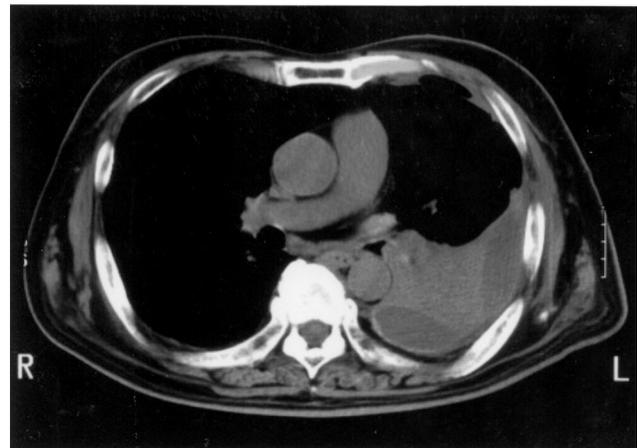


Fig. 3. The left lower lobar bronchus is obstructed by neoplastic growth; therefore the atelectatic left lower lobe is noted.

including concurrent benign-looking squamous cell elements (Fig. 4A), fusiform, and polygonal-shaped mesenchymal elements (Fig. 4B) in the fibromyxomatous stroma. Therefore a histological diagnosis of a pleomorphic adenoma was initially made. There was no evidence of a gradual transition between these elements. However, immunohistochemical staining demonstrated myoglobin-positive and smooth muscle actin (SMA)-negative rhabdomyosarcomatous elements (Fig. 5), in addition to cytokeratin-positive epithelial elements, so a final diagnosis of pulmonary carcinosarcoma was made.

The drainage catheter was removed on the 15th postoperative day, and the patient was discharged on the 46th postoperative day. He is now alive and doing well at 62 months following the surgical treatment, with no evidence of recurrence of the disease.

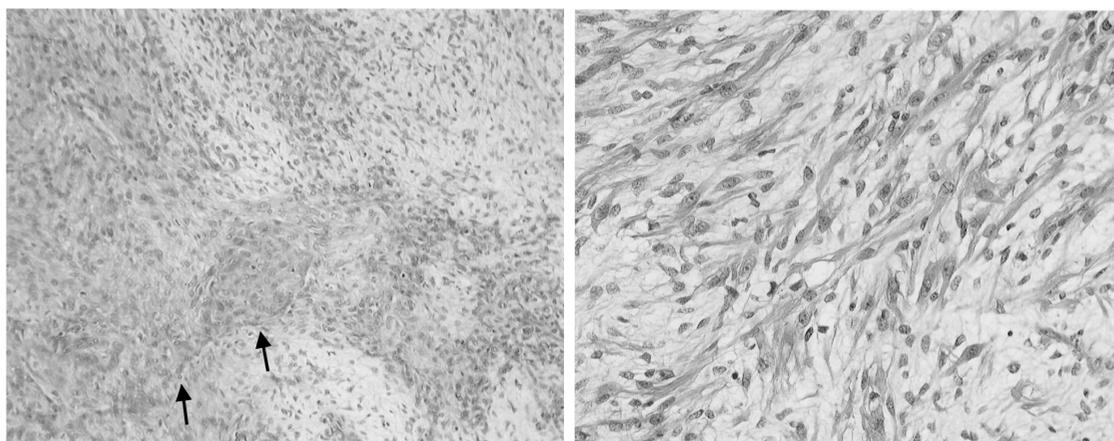


Fig. 4. The tumor composed of both squamous cell carcinoma elements and mesenchymal elements (hematoxylin and eosin staining).

A: Squamous cell elements (arrows).

B: Fusiform to polygonal mesenchymal cells.

Discussion

Pulmonary carcinosarcoma is a rare type of tumor. According to the World Health Organization's definition published in 1999, pulmonary carcinosarcoma normally presents as a malignant tumor that has a mixture of malignant epithelial and mesenchymal elements usually seen in adult malignancies.^{1,2)} It has been reported that carcinosarcoma accounts for from 0.2% or 0.27% of primary pulmonary malignancies. It is difficult to distinguish the tumor from spindle cell carcinoma unless a diagnostic differentiation is made regarding the specific tissues, such as neoplastic bone, cartilage, and striated muscle tissue.¹⁾ To ensure a correct pathological diagnosis, immunohistochemical techniques are recommended because they facilitate the differentiation between the epithelial and mesenchymal tumor elements.³⁾ The histogenesis of these tumors remains controversial. Most recently, Humphrey and associates suggested that these tumors were derived from a single stem cell, with the multipotentiality of lung tissue, thereby reflecting a spectrum of differentiation between carcinosarcoma and spindle cell carcinoma.⁴⁾

A pleomorphic adenoma of the lung is also a rare neoplasm. Pleomorphic adenomas are most frequently found in the salivary glands, but are well-recognized, though rare, bronchial neoplasms.⁵⁾ Most of these lesions tend to be located in the proximal major bronchi.⁵⁾ Bronchial glands are not usually found in the periphery of the lung.⁶⁾ Therefore the formation of the tumor in a major bronchus is consistent with the possible

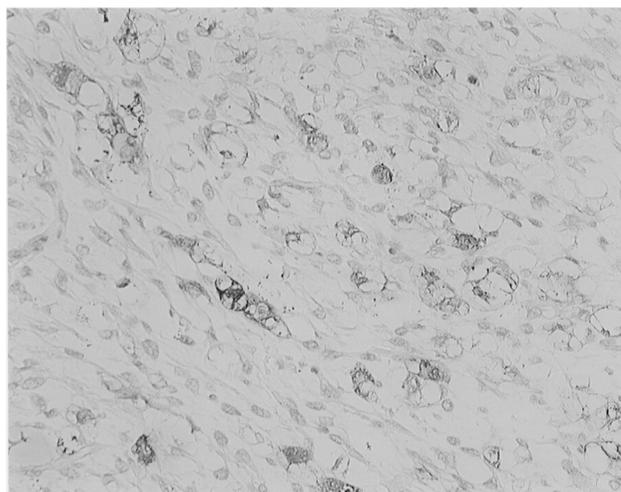


Fig. 5. Immunostaining for myoglobin confirmed the rhabdomyosarcomatous nature of the spindle-shaped mesenchymal elements.

origin from bronchial glands.⁵⁾ Pleomorphic adenoma has been shown to contain both epithelial and myoepithelial components.⁵⁾ Morphologic diversity is evident among different tumors and also within the same lesion. The epithelial component in a pleomorphic adenoma might form into ducts, cell nests, solid sheets, or interlacing cords. The mesenchymal stroma might be myxoid chondroid or fibrous.⁷⁾

Based on both the epithelial and mesenchymal components noted in the lesion and the common sites of these components, it was therefore necessary to differentiate between pulmonary carcinosarcoma and

pleomorphic adenoma in the present case.

Immunohistochemical staining for myoglobin showed rhabdomyosarcomatous elements, and therefore a diagnosis of true carcinosarcoma was made.

Moreover, pleomorphic carcinoma and pulmonary blastoma should also be considered in the differential diagnosis. However, a definite nonepithelial component such as a striated-muscle component can rule out the possibility of a pleomorphic carcinoma. Pulmonary blastoma can also be ruled out based on the absence of characteristic, glycogen-rich, juvenile, adenocarcinoma-like elements, in addition to the inconsistency regarding the patient's age and the site of tumor development.

Patients with either of these tumor types usually have a poor prognosis, despite undergoing such treatment modalities as surgery, radiation therapy, and chemotherapy.^{4,8)} The average postoperative survival time has been reported as 9 months, and fewer than 10% of patients survive for 2 years.⁹⁾ Several investigators have said that patients with central endobronchial tumors have a better prognosis than those with peripheral invasive tumors. However, this is observed only when the tumor is small (< 3 cm) and no metastases are present.¹⁰⁾

The current patient is now alive and well at 62 months following surgical treatment and has no evidence of recurrence of the disease. Although the surgical procedure was very difficult because of the presence of lung abscess in this patient, the outcome indicates that patients with carcinosarcoma do not always have an unfavorable prognosis. Therefore potentially curative surgical resections should always be attempted.

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