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Squamotous-type sarcomatoid carcinoma of the lung with rhabdomyosarcomatous components

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Abstract

Lung carcinosarcoma is an infrequently biphasic tumor composed of carcinomatous and sarcomatous components. It is divided into endobronchial (squamous-type) and peripheral (glandular type) categories. The carcinomatous component is usually a squamous carcinoma, and the sarcomatous component usually resembles a fibrosarcoma or a malignant fibrous histiocytoma. The presence of rhabdomyoblastic differentiation in such neoplasms is exceedingly rare. There are strong associations with smoking and asbestosis. In this study, we describe a unique case of a 43-year-old man with a 75 packet/year smoking history in whom a rare mixed malignant tumor of the lung was diagnosed and treated by left pneumonectomy. Histological examination of the resected specimen showed squamous cell carcinoma and rhabdomyosarcoma components. Although rare, the association of a sarcomatoid carcinoma of the lung with squamous cell carcinoma and rhabdomyosarcomatous component is possible and should be kept in mind when dealing with these unusual tumors.

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Full Text

Sarcomatoid carcinomas of the lung are a group of poorly differentiated non small cell carcinomas, mainly squamous cell carcinoma (45-70%) that contains a component of sarcoma or sarcoma-like differentiation. Carcinosarcoma is a mixture of carcinoma and sarcoma containing differentiated sarcomatous elements. The sarcomatous component is most often poorly differentiated spindle cell sarcoma. However, a careful search always shows areas of more specific sarcomatous differentiation, most often rhabdomyosarcoma. [1]

Although the development of carcinosarcoma is more closely associated with smoking, there are cases reported to be caused by asbestos exposure. [2],[3] We present a carcinosarcoma case in an active smoker with interesting macroscopic views. The longitudinal section of the postpneumonectomy pathology specimen extends to the left main bronchus and all lower subsegment bronchi in the shape of a glove finger, filling the airways without invading the wall. The underlined phrase does not make a sense.

A 43-year-old male had a smoking history of 75 packs/year and continued to smoke at the time of admission. He had started to suffer from dyspnea eight months ago and had lost 5 kg in the last 3 months. He presented at the hospital with increasing shortness of breath in the last few months and the chest x-ray revealed atelectasis in the left lung [Figure 1]a. Respiratory sounds were not heard on the left hemithorax on physical examination. A soft tissue appearance 53 × 18 × 15 mm in size and obliterating the left main bronchus, starting immediately from the carina in the lumen of left main bronchus was detected on computed tomography (CT) of the chest. In addition, there was a complete atelectasis of the left lung, and there were not pathological lymph nodes [Figure 1]b. Bronchoscopic biopsies were taken from the mass

located in the left main bronchus starting approximately 2 cm distally from carina. The pathology result was reported as malignant mesenchymal tumor. No other abnormal uptake besides the left lung mass, that showed hypermetabolic involvement and a maximum standardized uptake value (SUVmax) value of 7.4, was observed in the PET-CT scan. Cranial MRI was normal. The patient underwent a left thoracotomy. Pleural fluid was found and a sample was sent for intraoperative examination, which proved to be negative for malignant cells. Then, biopsies from lymph nodes in nodal stations 4L, 5, 7, 9, and 10 were performed, and were followed by a left pneumonectomy. At gross examination of the resected specimen, the left main bronchus was found to be filled with the mass, progressing to the lower lobe bronchus and subsegment bronchi but there was no macroscopic invasion of bronchial walls [Figure 2]a and b. The tumor was originated from lower lobe bronchial mucosa. The microscopic examination was diagnostic of sarcomatoid carcinoma (squamous cell carcinoma and rhabdomyosarcoma) and all lymph nodes were reported as reactive. The patient had an uneventful postoperative course and was placed into a chemotherapy program with (I fosfamid 5000mg/m², doxorubicine 40mg/m², vincristine 2, 3 mg/m²). One year after operation, there are no signs of recurrence. {Figure 1}{Figure 2}

Sarcomatous change in a carcinoma is a relatively rare event but has been reported by many investigators in various anatomic sites such as the skin, head and neck, thyroid gland, gastrointestinal tract, liver, gallbladder, pancreas, mammary gland, urinary tract, genital tract, and lung. [4] Sarcomatoid carcinomas of the lung are generally believed to be more aggressive and have a poorer prognosis than ordinary lung carcinomas. [5],[6] However, some studies have found sarcomatoid carcinomas not to be significantly more aggressive than ordinary lung carcinomas. [4] Fishback et al., report that cases beyond stage I, with lymph node involvement, a tumor size larger than 5 cm, with pleural invasion and the sarcomatoid component making up more than 50% of the tumor are associated with poor prognosis. [7] Our case was stage II, with 6 cm tumor size, with no pleural invasion or lymph node involvement, and the sarcomatoid component consisted of more than 50% of the tumor. The carcinoma extended from the main bronchus to the subsegments of the upper and lower lobes as finger-like extensions but did not invade the airways. We speculate whether the presence or absence of airway invasion could be a postoperative prognostic factor in addition to the criteria defined by Fishback et al. The patient is being followed-up and his condition will be closely monitored.

Smoking plays a primary role in the etiology of carcinosarcoma as in other lung cancers. Atypical and hyperplastic changes are histologically observed in the bronchial epithelium in 10% of smokers and atypical cells are observed at a rate as high as 96.7%. Squamous cell carcinomas metastasize later than other types, and usually tend to spread locally. [8] However, there may be necrotic and cystic areas in the rhabdomyosarcoma mass or it can extend and invade neighboring vessels and bronchi. Thorax localization is also reported to be a poor prognostic factor. [9] Our case was stage II, with a 6 cm tumor size, and the sarcomatoid part consisted of more than 50% of the tumor and with thorax localization, all poor prognostic factors. The absence of lymph node involvement and pleural invasion were good prognostic factors.

In a lung carcinosarcoma case published in 2001, actin and vimentin positivity were detected on staining in addition to the squamous cell carcinoma but the case was reported as carcinosarcoma as rhabdomyosarcoma or osteosarcoma could not be identified. [10] Our case was a squamous cell carcinoma with a moderately differentiated epithelial component. The majority of the sarcomatoid component was composed of rhabdomyosarcoma areas consisting of vimentin, skeletal muscle actin, and desmin positive spindle-shaped and rhabdoid cells.

Surgical resection of the lung tumor is the mainstay of the treatment but we administered chemo-radiotherapy to the patient as lung carcinosarcoma is rarely seen and still has no clearly identified treatment models and survival analysis.

In conclusion, although rare, the association of a sarcomatoid carcinoma of the lung with squamous cell carcinoma and rhabdomyosarcomatous component is possible and should be kept in mind when dealing with these unusual tumors.

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