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Malignant Fibrous Histiocytoma of the Lung

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ABSTRACT

Malignant Fibrous Histiocytoma (MFH) is the most common soft-tissue sarcoma which predominantly surfaces in the retro-peritoneum and extremity. MFH of lung is a very rare condition. The diagnosis is essentially through histologic examination. However, the invasion of the chest wall or other organs might help establish the malignant nature.We report a case of a 77-year-old man who had been presented with a history of dyspnea and non-massive hemoptysis. Computed Tomography (CT) scan of the chest showed a lobulated mass which had developed into the hilum of the lung. Furthermore, the inferior pulmonary artery was invaded. Thus, the final histological diagnosis confirmed MFH. The patient underwent a left pneumonectomy along with the dissection of the regional lymph nodes. Although lung MFH is rather an uncommon condition, it must be taken into consideration in differentiating between the types of lung tumor. What can be concluded is that complete surgical resection is the essential approach.

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Introduction

Malignant Fibrous Histocytoma (MFH) is the most common soft-tissue sarcoma which predominantly surfaces in the retro-peritoneum and extremity. MFH of the lung is quite a rare condition and the average of age for that is 55 years old. The ratio of developement in men to women is 30/23. Hemoptysis, weight loss, coughs and chest pain are the most prominent symptoms while Hypoglycemia and osteoarthropathy occur rarely. For the most part, the radiographic feature composes of a huge, solid mass in the lungs. Furthermore, calcification is seen infrequently. This tumor is often visible between the two lungs. Most of these types of tumors are pleomorphic and inflammatory while the myxoid type tends to occur rarely.The treatment consists of extensive and complete local excision, containing pulmonary and pleural resection followed by chemotherapy or radiotherapy. If the chest wall is involved, it requires resection. In patients with large tumors the use of neoadjuvant or postoperative chemotherapy is also advisable. The prognostic factors of the possible outcomes include the extension of the tumor into mediastinum or chest wall, extra thoracic metastasis and unsatisfactory excision. A survival period of approximately 5 years is reported in 15% of the cases (1).

Case

A 77-year-old man was presented with a four-month history of dyspnea and non-massive hemoptysis. His chest X-ray showed a mass of 5 cm, round and in the left hilar area of the lungs. Computed Tomography (CT) scan of the chest showed an approximately 5×4 cm lobulated mass arising anterior segmental bronchus in the leftupper lobe which was extending to the interlobar fissure, the lower lobe and the inferior pulmonary artery. Rigid bronchoscopic was done and the results of biopsy proved to be undifferentiated carcinoma (Figure 1).

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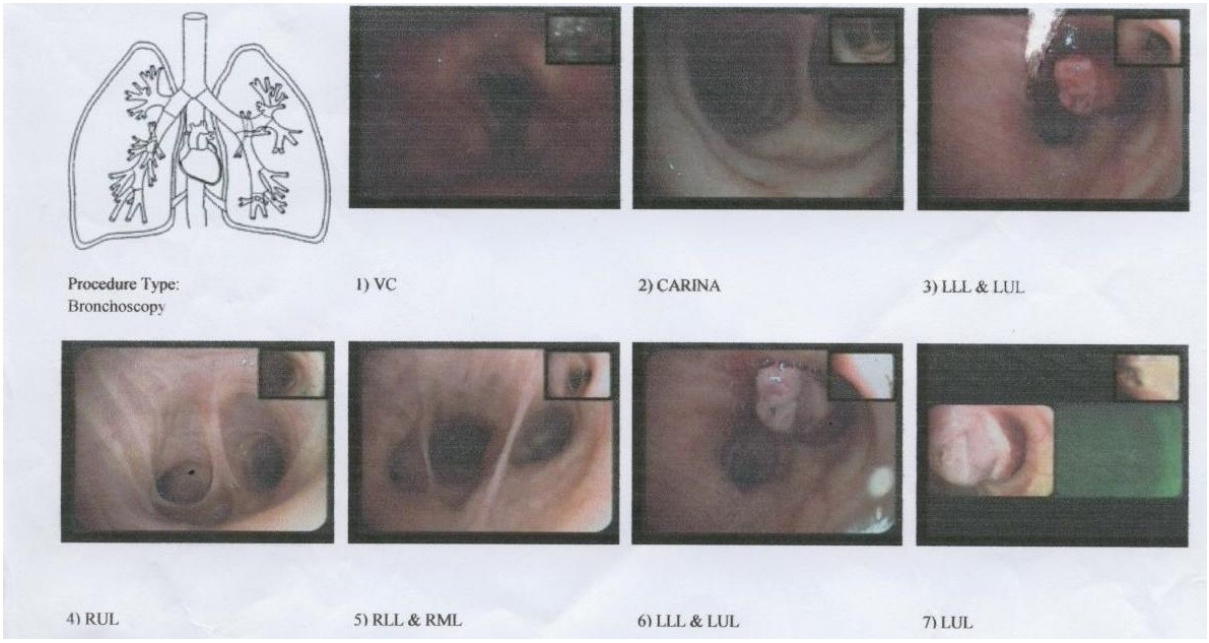

Figure 1. Bronchoscopy of protruded mass arising from superior segmental bronchus in lower lobe

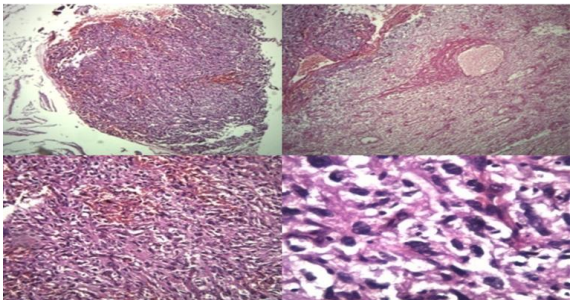

Figure 2. Microscopic feature consisting of psedustratified ciliated columnar epithelium with a mucinous gland diagnosed as typical Malignant Fibrous Histiocytoma

Pathology

The full appearance of the resected lung showed a tumor in the hilum of lung and it was firm with a size about 4*5 cm. The lower pulmonary arterial was also invaded by the tumor.

Microscopically, the tumor consisted of psedustratifiedciliated columnar epithelium with a mucinous gland. The nuclear cells had severe pleomorfism and acidophil cytoplasm without keratin perl accompanied with necrosis and extensive hemorrhage (Figure 2).

By an immunohistochemical staining, the tumor was positive for S-100 protein but negative for cytokratin (CK), desmin andepithelial membrane antigen (EMA). The final, histological diagnosis was typical MFH.

In the course of hospital admission, the patient did not seem to have any complications and thus, he was discharged after 7 days. The patient referred to the oncology ward later for chemotherapy. However, he did not go through any follow-up. As a result, we do not have the long-term reports on the late effects of the patient's surgery.

Discussion

MFH occurred most prominenetly in the extremity and retroperitoneum and was a soft-tissue sarcoma (2). Primary MFH is a very rare condition for the lungs (accounting for less than 0.2% of lung tumors). Nevertheless, metastasis to the lung from extra thoracic organ, also known as secondary MFH, is not as uncommon. Primary MFH could be diagnosed if there is no evidence of tumorous lesion - except for primary site- found through careful physical examination and para-clinical study (3). In our case, only pulmonary lesions appeared in the CT scan of the chest without extrathoracic lesion.

Maeda et al. searched English Medical Literature in the world up until 2007 and recorded 93 patients diagnosed with MFH (4). The patients were aged between 18 - 80 years and the mean age was 55 years. The most common clinical symptoms were chest pain, cough, hemoptysis, and weight loss. Hypertrophic pulmonary osteoarthropathy and hypoglycemia were also observed in a few patients (1). In our case, a 77-year-old man had such clinical symptoms as dyspnea and non-massive hemoptysis.

The tumor had appeared in the CT scan and since he had been presented with non-massive hemoptysis, we performed rigid broncoscopy. The result of pathology was undifferentiated carcinoma. Maeda et al. claimed that a preoperative diagnosis is not easy and only 4% of their reported patients had been histologically diagnosed prior to resection. The rarity of MFH as well as inadequate biopsy specimen might be the reason (4).

Many differential diagnoses have been made

for Malignant Fibrous Histiocytoma; for instance, malignant schwannoma, spindle-cell carcinoma, other types of sarcoma, anaplastic carcinoma and malignant melanoma. It should be noted that immunohistochemical stain is essential for an accurate diagnosis in this case. Many variants include angiomatoid, giant cell, myxoid, pleomorphic and storiform observed in the immunohistochemical stain (3). In our case, microscopically, the tumor consisted of pseudstratified ciliated columnar epithelium along with a mucinous gland. The nuclear cells had severe pleomorfism and acidophil cytoplasm without keratin perl which were accompanied with necrosis and extensive hemorrhage. By an immunohistochemical staining, the tumor was positive for S-100 protein and negative for cytokratin (CK), desmin and EMA. The final, histological diagnosis was typical MFH.

Huang C et al. reported the clinical information and survival status of 20 patients with primary Malignant Fibrous Histiocytoma of the lungs.The clinical symptoms were cough, non-massive hemoptysis, chest pain, fever and chest discomfort. The 1-year and 2-year overall survival rates were 55.0% and 25.0% respectively. Unfortunately, the majority of the patients died of local recurrence and distant metastasis. Patients with tumors smaller than 5 cm had a survival period of 27 months. On the other hand, those with larger tumors than 5 cm survived for a period of 8 months. The median survival of the central tumors was 6 months. However, the median was about 23 months for the peripheral tumors. The most effective managing process proved to be surgery and the role of postoperative adjuvant chemotherapy was indeterminate. It is also noteworthy that small mass size and peripheral type in lung tumors might be associated with a relatively improved prognosis (5).

Maeda J et al. recommended complete resection with the systematic dissection of regional lymph nodes for exerting a better effect in the survival of patients with primary pulmonary MFH (4).

Conclusion

We reported a case of primary MFH of the lung who was treated during a surgical process successfully. Although this kind of diagnosis is quite rare, it should be considered in the differential diagnosis of lung tumors.

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Conflict of interests

The author has no conflict of interests.

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