A Case Report: Primary Malignant Fibrous Histiocytoma of the Lung

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Abstract

Primary malignant fibrous histiocytoma (MFH) of the lung is very rare and only a few cases have been reported in the literature. A 69 year-old man presented with hemoptysis and dyspnea of three months duration. Thoracic computed tomography (CT) scans revealed a lobulated mass invading the mediastinum. Histopathologic examination of bronchoscopic biopsy revealed a stroiform-pleomorphic pattern of MFH. As no abnormal lesions were detected in the abdomen and extremities, the tumor was diagnosed as a primary MFH of the lung.

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Key words: *primary malignant fibrous histiocytoma of the lung, thoracic computed tomography*

Introduction

Malignant fibrous histiocytoma (MFH) is a primitive sarcoma originating in the deep soft tissues and characterized by a bimorphic population of fibrocytic and histiocytic cells usually arranged in a storiform pattern (1-3). Although it is one of the most common soft tissue sarcomas of adulthood accounting for about ten percent of all sarcomas, the lung is an extremely uncommon primary origin for the tumor (4-5).

Case Report

A 69 year-old man presented with hemoptysis and dyspnea of three months duration. The patient had been rather well in the past. He was a farmer and had been smoking two packs of cigarettes daily for 55 years. The physical examination at the time of admission did not disclose any abnormality. Complete blood count and serum biochemistry were within normal limits. Chest x-ray showed a mass lesion in the mid-right lung zone and also widening of upper right side of mediastinum (Figure 1). Thoracic CT scans revealed a mass invading the mediastinum (Figure 2). Bronchoscopy revealed tumoral infiltration of distal one-third of trachea and also right main bronchus, and biopsies were done. Histopathologic examination of the biopsies was consistent with storiform-pleomorphic pattern of MFH

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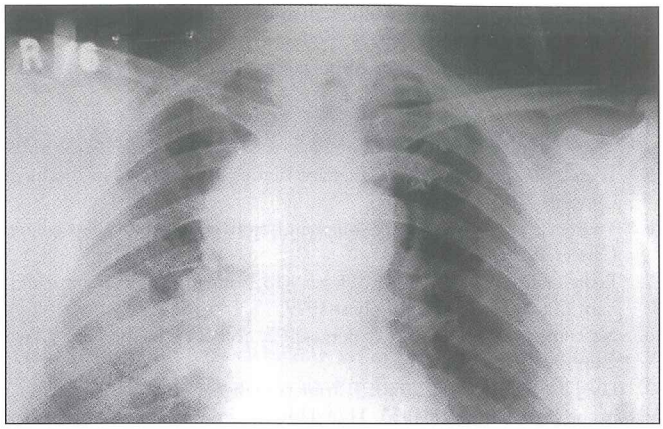

Figure 1. *Posteroanterior chest x-ray showed a mass lesion in the mid-right lung zone and also widening of upper right side of mediastinum*

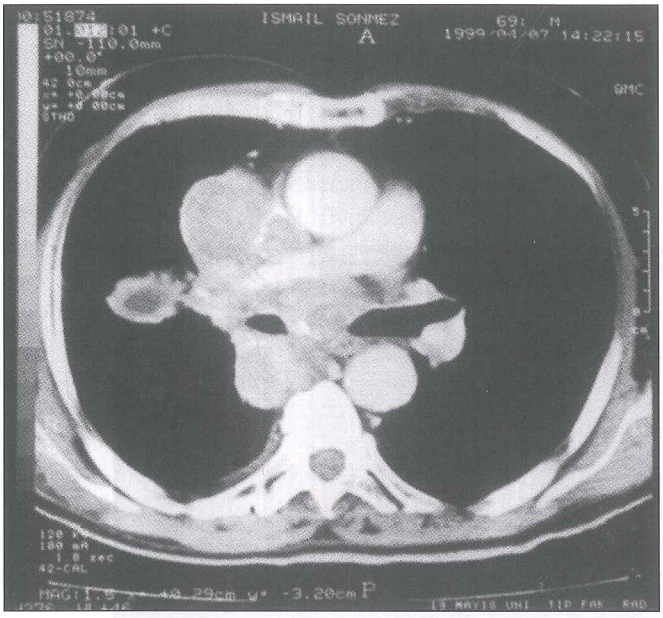

Figure 2. *Thoracic CT scan at two cm caudal to the tracheal carina demonstrated a bulky right hilar mass encasing the right pulmonary artery and the superior vena cava*

(Figure 3). And also, tumor cells showed positive staining for vimentine and negative for cytokeratin.

The staging procedures were done to determine whether the tumor was primary or metastatic, and if primary, whether it was resectable or not. The ultrasonography and CT of the abdomen, magnetic resonance imaging of both extremities, bone scintigraphy and cranial CT were all normal.

Thus, the final diagnosis was primary MFH of the lung extending into the mediastinum and nearby vascular structures, without metastasing beyond the thorax. Due to the involvement of the mediastinum, the tumor was regarded as unresectable and radiotherapy of 5000cGy was applied, but no response was seen at the end of radiotherapy. The patient died 17 days after radiotherapy.

Discussion

Malignant fibrous histiocytoma is among the most common soft tissue sarcomas of adulthood, but rarely originates from the lungs (4-5). Its etiology is unknown. Mean age is 52 years and there is a slight preponderance of females. Clinically, the patients usually complain of chest pain, cough and hemoptysis. Chest x-ray usually shows solitary mass lesion, but there are reports of more than one mass lesion in the literature (6-7). In our case, there was solitary pulmonary lesion at mid-right lung zone and also widening of upper right side of mediastinum. Thorax CT, in our case, demonstrated a lobulated mass invading the mediastinum. Nascumento et al. (8), found ten percent of sarcomas of the lung to have a dominant endobronchial component. Our case had also an endobronchial component.

Histopathologically, four types of MFH are described, of which the most common one is storiform-pleomorphic type (5-6), as in our case. Also tumor cells showed positive staining for vimentine and negative for cytokeratin that were compatible with MFH (6). Since primary MFH of the lung is very rarely reported, if diagnosis of MFH is reached histopathologically, a comprehensive evaluation should be immediately made to show whether it is metastatic, usually originating from lower extremities and abdomen, or primary.

Only after the evaluation has been found completely normal in other organs, the diagnosis of primary MFH of the lung could be reached. We have found no abnormalities related to its possible primary sites other than the lungs at the end of the evaluation, and we concluded that it was a primary MFH of the lung.

Invasion of mediastinum and chest wall at initial diagnosis, and recurrence or metastasis are poor signs (9). However, pleural invasion, size, peripheral or central location in the lungs, and mitoses or necrosis are not significant predictors of future behaviour.

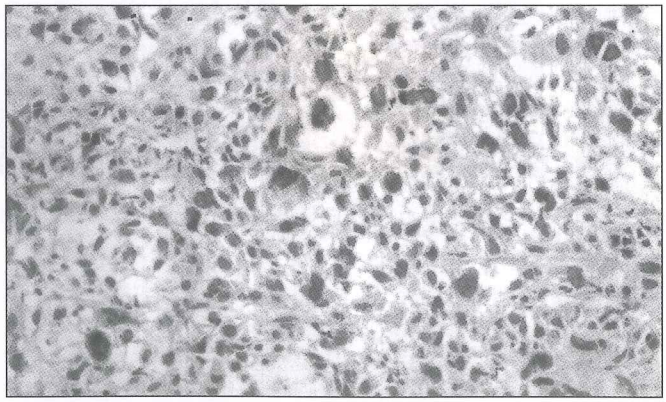

Figure 3. *Histopathological examination of the biopsies disclosed a storiform-pleomorphic pattern of MFH*

Surgery is the primary mode of therapy, with high rates of local and distant recurrences reported (10), but the role of radiation therapy and chemotherapy has not been clearly defined. In our case, due to mediastinal involvement, the tumor was unresectable and radiotherapy was chosen as a primary therapeutic modality, but the tumor did not respond to the treatment.

In conclusion, although primary MFH of the lung is among the extremely rare tumors of the lung, it should be considered in differential diagnosis of pulmonary masses.

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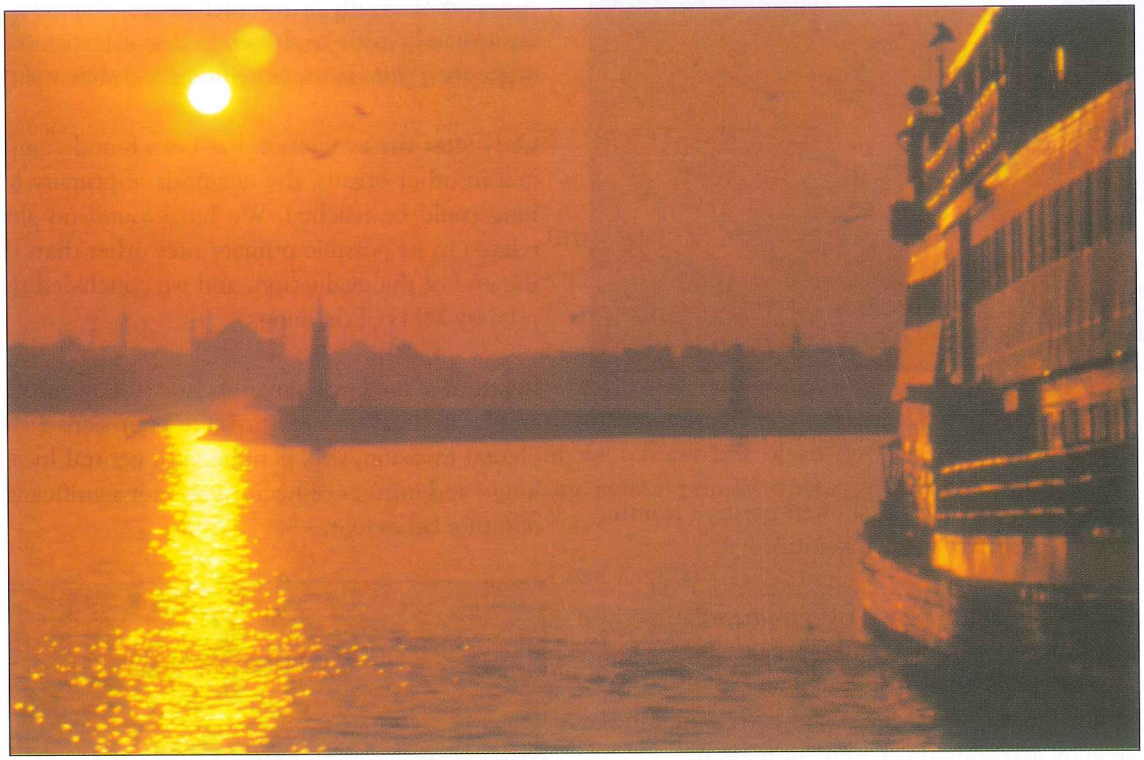

A view from Bosphorus; Photography by Kamil Levent Arslan, MD