Journal of BUON **14:** 143-145, 2009

Primary myofibrosarcoma of the lung. A rare case with challenging diagnosis

A. Lioulias[1], P. Misthos[1], K. Neofotistos[1], S. Legaki[2]

[1]Thoracic Surgery Department and [2]Pathology Department, "Sismanogleio" General Hospital, Athens, Greece

Summary

Primary myofibrosarcoma (MFS), or myofibroblastic sarcoma of the lung, is a rare tumor. A 47-year-old man with a history of melanoma presented with a central tumor of the left lung. The initial diagnosis was inflammatory myofibroblastic tumor but after extensive pathologic analysis MFS of the lung was confirmed. He underwent left pneumonectomy and died on the 6th postoperative month. This is the second case of primary pulmonary MFS to be reported.

Key words: histology, lung, myofibrosarcoma

Introduction

Primary MFS of the lung is an extremely rare lung tumor. Diagnosis demands extended histopathological analysis, making differential diagnosis and decision of definite management very difficult. This is the second case of primary MFS of the lung to be reported [1].

Case presentation

A 47-year-old man with a history of right arm melanoma, which was surgically managed 8 years ago, was on regular follow-up. On April 9, 2006 a thoracic CT scan showed a central solid homogeneous mass in the left lung (Figure 1). Physical examination was unremarkable and the patient had no history of trauma or any kind of previous surgical procedure. Bronchoscopy showed that the bronchial mucosal surface was normal with signs of mild external pressure at the level of secondary carina. Transbronchial needle aspiration was inconclusive for the nature of the tumor.

A left lateral thoracotomy was performed. Multiple frozen sections from the tumor showed no malignancy and suggested inflammatory tumor of the lung. However, the infiltrative nature of the tumor raised a strong suspicion for the malignant nature of the lesion. Since the only radical resection for that young person was left intrapericardial pneumonectomy, it was decided to wait until the final histological results, which were indicative of lung sarcoma. The patient returned to the operating room and was subjected to left intrapericardial pneumonectomy with en bloc partial left atrial resection. The histological results of the surgical specimen confirmed the diagnosis of MFS.

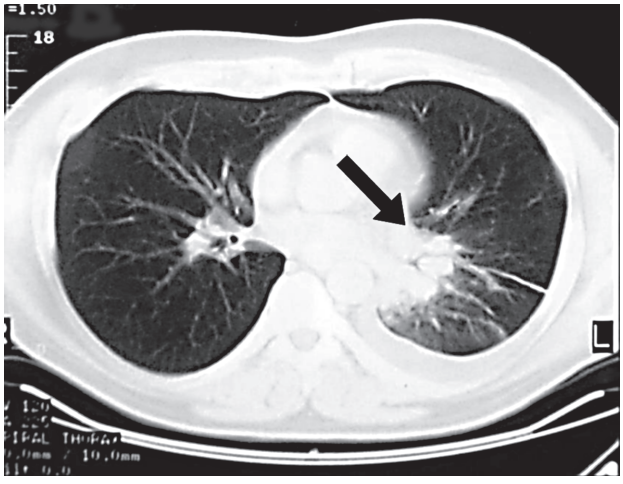

Figure 1. Computed tomographic scan of the thorax: central left lung solid tumor (arrow).

Correspondence to: Panagiotis Misthos, MD. 16-18 Markou Avgeri Street, 15343 Agia Paraskevi, Athens, Greece. Tel: +30 210 6080107, Fax: +30 210 6080107, E-mail: panmisthos@yahoo.gr

Received 24-04-2008; Accepted 10-07-2008

The tumor had a quite uniformly cellular spindle cell proliferation comprising elongated cells that exhibited a mild to moderate degree of nuclear pleomorphism, and lying in loose fibrous stroma. There was minimal pleomorphism without necrosis, and the mitotic index was 16 per 10 hpf in some areas. Few, predominantly chronic inflammatory cells associated with the tumor were evident, although they were not particularly numerous (Figure 2). There was focal inflammation which was mostly lymphocytic but with occasional plasma cells. The tumor was diffusely infiltrating the lung parenchyma, and extensively involved the pulmonary vessels (Figure 3). Immunohistochemistry showed strong staining of spindle cells for smooth muscle actin (SMA), and negative staining for desmin, MNF116, S100, EMA, ALK1, melan A and CD3, 20, 30, 34.

The postoperative course was uneventful and the patient was discharged on the 12th postoperative day. The patient died 6 months later due to pulmonary embolism.

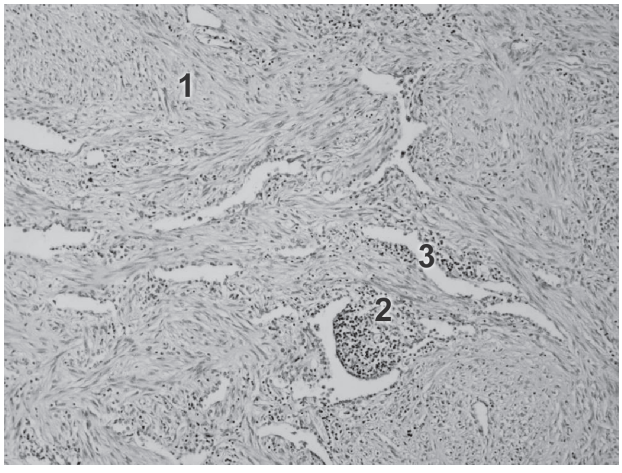

Figure 2. The tumor is quite uniformly fascicular (1) and there are scattered inflammatory cells (2). Alveolar infiltration (3) (H&E ×40).

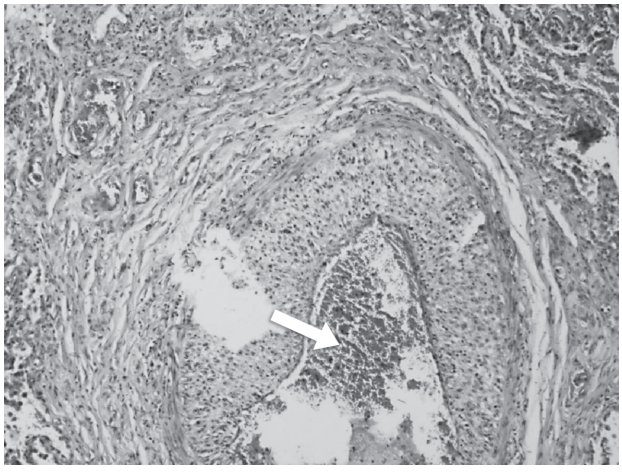

Figure 3. Vascular invasion by the tumor (arrow) (H&E ×100).

Discussion

Primary MFS, or myofibroblastic sarcoma, is a rare tumor of the lung. To our knowledge this is the second case to be reported [1]. The authors encountered a serious problem of differential diagnosis between inflammatory tumor of the lung (benign condition) and pulmonary MFS (aggressive malignant condition). Furthermore, it was imperative to exclude the possibility of melanoma metastasis. We did not proceed outright to left pneumonectomy during the first operation because in case of an inflammatory disease it would be valuable to attempt shrinkage of the inflammatory tumor with corticosteroids or other medication in order to achieve radical resection with lesser operation, such as lobectomy or even sleeve lobectomy. The use of corticosteroids has been proposed for functionally inoperable patients, those with unresectable lesions and with disease relapse [2].

A variety of differential diagnoses had to be considered. Firstly, melanoma metastasis was excluded, apart from the atypical morphology, by the negative immunostains for S-100 protein and melan A. Inflammatory myofibroblastic tumor, sarcomatoid mesothelioma, lymphoma, leiomyosarcoma, fibrosarcoma, and lung carcinoma were excluded by the pathological study that showed long spindle cells with the immunophenotype of myofibroblasts (positive for SMA, and negative for desmin, MNF116, S100, EMA, ALK1, melan A and CD3, 20, 30, 34). Especially for inflammatory myofibroblastic tumor of the lung the lesion lacks the typical mixture of histiocytes, lymphocytes, fibroblasts, and, in particular, plasma cells, with the fibroblasts showing positive expression of ALK [3]. Plasma cells were present in our case but not as the predominant cell population. Moreover, the tumor was quite uniformly fascicular with no much background fibrosis and inflammation. Even if this was an inflammatory myofibroblastic tumor, the neoplasm showed a strikingly infiltrative pattern with important vascular invasion. Based on the focal nature of the inflammation, we thought proper to considered it as MFS (grade 1-2, according to the French grading system).

Sarcomatous transformation of both pulmonary and extrapulmonary inflammatory myofibroblastic tumor has been described in previous reports. We found 5 such cases in the literature [4-6], 2 of them in the lung. Usually, sarcomatous transformation takes a long period of time and is usually detected after several recurrences of the inflammatory tumor. No such events were encountered in our case. The previous CT scan was done 18 months before the diagnosis and was negative.

In conclusion, any infiltrative lung tumor with the characteristics of inflammatory tumor should be examined for the possibility of MFS, since inflammatory pseudotumors may histologically resemble low-grade sarcomas or focally transformed to MFS. Frozen section biopsies were misleading. The authors recommend that extensive pathological analysis should herald any decision for definite management.

References

1. Morawietz L, Kuhnen C, Katenkamp D, Le Coutre P, Ladhoff A, Petersen I. Unusual sarcomatoid neoplasm of the lung suggesting a myofibrosarcoma. Virchows Arch 2005; 447: 990-995.
2. Urschel JD, Horan TA, Unruh HW. Plasma cell granuloma of the lung. J Thorac Cardiovasc Surg 1992; 104: 870-875.
3. Melloni G, Carretta A, Ciriaco P et al. Inflammatory pseudotumor of the lung in adults. Ann Thorac Surg 2005; 79: 426-432.
4. Spencer H. The pulmonary plasma cell/histiocytoma complex. Histopathology 1984; 8: 903-916.
5. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol 1995; 19: 859-872.
6. Donner LR, Trompler RA, White RR. Progression of inflammatory myofibroblastic tumor (inflammatory pseudotumor) of soft tissue into sarcoma after several recurrences. Hum Pathol 1996; 27: 1095-1098.