Case report

Obstructing endobronchial malignant fibrous histiocytoma

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Received 6 June 2000; received in revised form 26 January 2001; accepted 14 February 2001

Abstract

A 58-year-old male patient admitted with malignant fibrous histiocytoma of the left mainstem. Rareness and unusual presentation of malignant fibrous histiocytoma and surgical management for lung salvage made its reporting worthwile. In this localization, prognostic characteristics are better than in the other localizations of lung and body. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Malignant fibrous histiocytoma; Soft tissue sarcoma; Endobronchial tumor

1. Introduction

Malignant fibrous histiocytoma (MFH) is the most common sarcoma of soft tissue in adults [1]. Primary pulmonary MFH is reported extremely rare in endobronchial location [1,2]. We report a case of primary pulmonary MFH that caused total atelectasis of left lung. In this report, its clinic, method of diagnosis, differential histological diagnosis and current approach to treatment are discussed.

2. Case

A 58-year-old male patient was admitted with cough, dyspnea and left chest pain. He had fever and sweating approximately 3 months before the admission, cough and production of sputum followed these. Then he noted left chest pain and 1 month later he admitted to an outside hospital where chest roentgenogram showed total atelectasis of left lung. Therefore, the patient referred to our hospital for further evaluation. In the physical examination dullness, decreased fremitus and diminished breath sounds were found in the left hemithorax. Chest roentgenogram showed indirect signs of total atelectasis (Fig. 1). Computerized tomography of chest confirmed a tumor, which was obstructing the left mainstem completely and placed into 3 cm distal of the carina. The bronchoscope could not be passed beyond the tumor and no attempt made to perform a biopsy with the fear of bleeding. Thoracotomy performed to the patient and the lung was atelectatic, edematous and filled with pus. A longitudinal bronchotomy performed just distal to the tumor. The tumor sized 2 × 2 cm and completely excised with a portion of bronchial wall, distal of the bronchus lavaged. Total lung expansion obtained after the operation. The bronchus closed with interrupted nonabsorbable sutures. Microscopic examination of tumor revealed evident cell pleomorphism and histiocytic elements, which were showing frequently atypical mitosis and rarely giant cell character. In the ground inflammatory cells were seen and evident phagocytosis was observed (Fig. 2). Also the margin of bronchial wall was without tumor. One year later, follow-up bronchoscopy showed no evidence of recurrence. His chest roentgenograms and pulmonary functions returned to normal ranges. And when the patient checked up 3 years later after the operation, he remained well and bronchoscopy showed no evidence of recurrence.

3. Discussion

MFH is a mesenchymal tumor first recognized by O’Brien and Stout in 1964 [3]. It is now considered to be the most common malignant soft tissue tumor of adults [1,4,5]. Because of the ubiquitous nature of mesenchymal tissue,
MFH could be seen in all organs. The most frequent location of MFH in soft tissues with a higher incidence are deep musculature of the extremities, retroperitoneum and trunk. The second most prevalent tissue for MFH is long bones and metaphyseal areas [6], but it is also reported in the lung. Primary pulmonary MFH is extremely rare, however, the lung is the most frequent involved organ by metastatic MFH [1,4,5].

The frequency of primary sarcomas of the lung is one case per 500 pulmonary sarcomas. The most common types are fibrosarcomas and leiomyosarcomas, MFH follows these in incidence [2]. MFH of the lung is first reported by Kern et al. in 1979 [7]. They usually arise from distal parenchyma and rarely are associated with a major bronchus, but occasionally are seen as a small endobronchial polypoid lesion [1,2]. Nascimento et al. [8] found that 10% of sarcomas of the lung are having a dominant endobronchial component. Yousem et al. [1] reported two cases of MFH from 22 cases in this location. Our case is an example for obstructing endobronchial MFH.

Clinical specialities, incidence and sex distribution are same as extrapulmonary MFH. The tumor can occur in all ages, but frequently in fifth and sixth decades [6,8]. Symptoms are non-specific and some patients are asymptomatic. Chest pain, cough, weight-loss, hemoptysis and dyspnea are the most prominent features. Two pulmonary MFH cases who had histories of asbestos exposure have been reported [5]. Also hypoglicemia is reported with MFH [6,8]. In our case hypoglycaemia was not found.

Although radiological appearance of the tumor in the lung is non-specific for MFH, two different patterns are reported: Solitary pulmonary nodule and diffuse infiltrate. Pulmonary MFH usually shows a peripheral uncavitated mass lesion without calcification [1]. Yousem et al. [1] reported 14 cases in peripheral location, four cases in central location and two cases in endobronchial in his series. However, Gilman et al. [9] reported a cavitary lung metastases from primary MFH of duodenum. Chest and abdominal computerized tomographies are helpful in delineating involvement of mediastinum, detecting subdiaphragmatic extension, and/or identifying primary retroperitoneal tumor. In CT examination, MFH has a predominantly muscle-density that is homogeneous in 45% of cases and heterogeneous in 55%. Increased uptake of radionuclides by the tumor can be observed in perfusion scanning examination. Angiographically, MFH shows hypervascularity and early venous return.

MFH has no specific clinical and radiological features for distinguishing from more frequently seen epithelial tumor [1]. Early diagnosis of MFH obtains a better prognosis because of the invasion to mediastinum and chest wall, recurrence and metastasis in advanced stages. MFH is rarely diagnosed with metastatic lesions before the discovery of primary source [6]. Indeed, MFH has a high tendency for local invasion in soft tissue, as well as for distant metastases, especially to the lung. Weiss and Enzinger [6] noted local invasion in 41% of cases, metastases in 42 and 82% of all metastases were to the lung. The metastatic pattern of primary pulmonary MFH reflects the high incidence of vascular invasion (50% of cases) and metastases are usually seen in brain, liver and lung [1]. Therefore, pulmonary MFH must be considered as a high grade malignant tumor. However, Weiss and Enzinger [6] were reported only one case from a series of 200 cases that had a pulmonary lesion before the discovery of primary source.

References


