

PRIMARY PULMONARY BIPHASIC SYNOVIAL SARCOMA: A CASE REPORT

PRİMER PULMONER BİFAZİK SİNOVİYAL SARKOM: OLGU SUNUMU

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Özet

Primer pulmoner sinoviyal sarkom nadir görülen bir akciğer tümörüdür. Bu yazıda öksürük ve nefes darlığı yakınmaları ile başvuran yaşlı bir olgu değerlendirildi. Pozitron emisyon tomografi ile primer lezyon tarandıktan sonra plevra biyopsisi ile tanı alan hasta inoperabl olduğu için doksorubisin/ifosfamid kemoterapisi uygulandı. (Anatol J Clin Investig 2009;3(4);235-238).

Abstract

Primary pulmonary synovial sarcoma is a rare lung tumour. In this study we present an elderly female with complaints of cough and dyspnea. Patient's primary lesion was evaluated with positron emission tomography - computed tomography. Pleural biopsy sample was taken and as the patient was inoperable, doxorubicin/ifosfamide chemotherapy was initiated. (Anatol J Clin Investig 2009;3(4);235-238).

Introduction

Synovial sarcomas are tumors originating from periarticular areas of the joint. These tumors mostly involve the lower extremities (50%) around the knee region. Rarely, they can be conferred in regions not associated with synovial structures, such as head-neck region (10%), thoracic and intraabdominal walls (10%) [1,2]. Primary pulmonary sarcomas account for less than 0.5% of all pulmonary malignancies [3]. The tumor is referred as biphasic due to its involvement of two different morphologic cell types. The aim of the present study was to report a synovial sarcoma case which is rarely encountered in literature.

Case

A 76-year-old female presented with complaints of cough and dyspnea increasing over the last two months. There was no history of cigarette smoking and alcohol use. Her personal history was unremarkable; but in her family history, her sister had lung cancer and her son had prostate cancer. Her vital signs were normal. Physical examination revealed decreased breath sounds at the base of the left lung and the presence of rales in the middle zone of the left lung. Posteroanterior (P-A) chest x-ray of the patient revealed pleural thickening, pleural effusion and parenchymal nodules in the left lung. Biochemistry analysis and hemogram were normal. Pleural effusion material obtained during thoracentesis was consistent with chronic inflammation. Polymerase chain (PCR) reaction for *Mycobacterium Tuberculosis* was negative

and there was no growth in the culture in the pleural effusion sample. The patient's positron emission tomography - computed tomography (PET-CT) demonstrated markedly increased F-18 fluorodeoxyglucose (FDG; Maximum standardized uptake value, SUVmax: 5.5) uptake in the interlobar fissure of all pleural areas of the left lung (Figure 1), a hypermetabolic lesion (SUVmax: 5.9) with pleural invasion in the apicoposterior segment of the left lung (Figure 2), two hypermetabolic lesions, with one invading the pleura, in the superior segment of the left lower lobe (SUVmax: 5.4 and 5.6, respectively) (Figure 3 and 4 respectively), a left parasternal lymph node (SUVmax: 4.5) (Figure 3), and a bronchopulmonary lymph node in the lateral basal segment of the left lower lobe (SUVmax: 6).

Malignant tumor proliferation demonstrating infiltrations in the form of solid areas among the necrotic areas, as well as atypical mitosis was observed in the pleural biopsy samples. Immunohistochemical analysis of the tumor cells were as follows: vimentin (+), S-100 (+), pancytokeratin (+), HMW-CK (+), CD99 (+), bcl-2 (+), isolated CD10 (+), EMA (-), CEA (-), smooth muscle actin (-), desmin (-), HMB-45 (-), CD34 (-), CD141 (-), RCC (-), calretinin (+) in mesothelial cells, and cytokeratin 5/6 (+) in reactive mesothelial cells, and the patient was diagnosed as biphasic synovial sarcoma. As the patient was inoperable, doxorubicin/ifosfamide chemotherapy was initiated.

Discussion

Synovial sarcoma is a mesenchymal neoplasm which accounts for 10% of primary malignant soft tissue tumors [4]. It primarily originates from periarticular soft tissues of the extremities. Currently, synovial sarcoma is considered to be originated from pluripotent mesenchymal cells presenting epithelial differentiation [5]. These regions include the head-neck region, mediastinum, heart, esophagus and vulva [6]. Primary pulmonary sarcomas account for less than 0.5% of all lung tumors and the most commonly encountered ones are leiomyosarcoma, malignant fibrous histiosarcoma (MFH), fibrosarcoma and synovial sarcoma [7].

The most frequent complaints of patients diagnosed with primary pulmonary sarcoma are chest pain, dyspnea and cough. Overall, 24% of patients are asymptomatic and diagnosis is made by an incidental lesion on chest x-ray. Our present case also admitted to our clinic with complaints of cough and dyspnea.

Following identification of the lesion by P-A chest x-ray and thoracic tomography, extrapulmonary involvement was evaluated by PET-CT. Extramediastinal involvement was not observed; however, hypermetabolic areas were found in the pleura and parenchyma of the left lung.

Although magnetic resonance imaging (MRI) is traditionally used in the evaluation of soft tissue tumors, increased F-18 FDG uptake in PET-CT describes the rarely encountered soft tissue tumors much better. While MRI does not clearly distinguish the fibrotic tissue, inflammation and recurrence which develops after surgery, PET-CT enlightens the clinician on this issue [8].

The basic treatment approach in these kinds of tumors is the extensive surgical resection. The most important prognostic factor is the complete resection of the tumor [9]. Even though severity of tumor only pleural biopsy was taken and complete resection could not performed in our case. Radiotherapy is recommended in cases with postoperative positive margin [10]. Doxorubicin and ifosfamide chemotherapeutic regimens may be administered as adjuvant treatment and at recurrence (11). Though the prognosis was limited to a few months previously; it has been prolonged to over a year with current multimodal treatment approaches [11].

In conclusion, following the elimination of extramediastinal lesions by PET-CT, an elderly case who admitted with the complaints of cough and dyspnea to our clinic, diagnosed as pulmonary synovial sarcoma by pleural biopsy and treated by chemotherapy was presented.



Figure 1. Patient's positron emission tomography - computed tomography demonstrating markedly increased F-18 FDG (SUVmax: 5.5) uptake in the interlobar fissure of all pleural areas of the left lung.

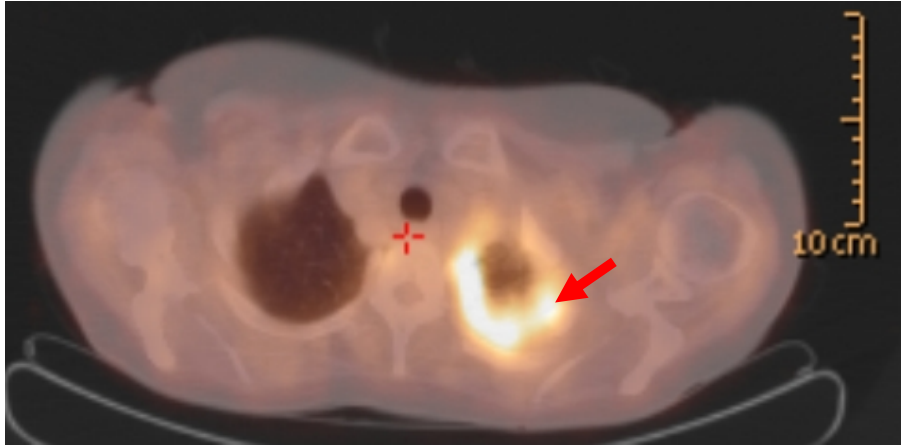


Figure 2. Patient's positron emission tomography - computed tomography demonstrating a hypermetabolic lesion (SUVmax: 5.9) with pleural invasion in the apicoposterior segment of the left lung.



Figure 3. Patient's positron emission tomography - computed tomography demonstrating hypermetabolic lesion invading the pleura in the superior segment of the left lower lobe (SUVmax: 5.4) and a hypermetabolic left parasternal lymph node (SUVmax: 4.5).



Figure 4. Patient's positron emission tomography - computed tomography demonstrating a hypermetabolic lesion in the superior segment of the left lower lobe (SUVmax: 5.6).

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