An old woman with weight loss and chest pain

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ÖZET

Göğüs ağrısı ve kilo kaybı ile başvuran yaşlı kadın hasta

Pulmoner sarkomlar, tüm primer akciğer sarkomalarının sadece %0.1-0.5'ini oluşturur. Bu tümörler akciğer parankimi, bronş ağacı veya pulmoner arterlerden köken alabilir. Ayırıcı tanıda en önemli nokta metastatik sinovyal sarkom ile ayırıcı tanısını yapmaktır. Yetmiş altı yaşında kadın hasta, son bir aydır süregiden ateş, öksürük, balgam, kilo kaybı ve sol yan ağrısı ile başvurdu. Toraks tomografisinde genişlemiş mediastinal lenf nodları ve sol plevral efüzyon saptandı. Torasentezinde hemorajik görünümlü, lenfosit hakimiyetli eksüda karakterinde sıvı saptandı ve kapalı plevra biyopsisi kronik inflamasyon gösterdi. Lokal anestezi ile yapılan sol torakoskopisinde sol akciğerde total kollaps ve viseral plevrada çok sayıda nodüller görüldü ve bu nodüllerden çok sayıda biyopsiler alındı. Patolojik tanısı "sinovyal sarkom" olarak kondu. İskelet single foton emisyon tomografisinde başka bir lezyon saptanmadı ve hasta primer plöropulmoner sinovyal sarkom olarak kabul edilerek kemoterapisi planlandı. Sonuç olarak, primer plöropulmoner sinovyal sarkom akciğer ve plevranın nadir rastlanan ancak prognozu kötü olan bir neoplazisidir.

Anahtar Kelimeler: *Plöropulmoner sarkom, torakoskopi.*

SUMMARY

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Pulmonary sarcomas constitute only 0.1-0.5% of all primary lung malignancies. These tumors may derive from the lung parenchyma, bronchial tree or pulmonary arteries. The most important entity in the differential diagnosis is metastatic synovial sarcoma. A 76-years-old woman was admitted for investigation of a fever, productive cough, dyspnea, weight loss and left-sided chest pain which had been present for one month. A chest computerised tomography showed enlarged mediastinal lymph nodes were observed, as well as a left-sided pleural effusion. Thoracentesis revealed hemorrhagic pleural effusion which was exudate and lymphocyte predominant, closed pleural biopsy showed chronic inflammation. Left sided thoracoscopy was performed under local anesthesia, total collapse of left lung and multiple pleural nodules were observed on the visceral pleura multiple biopsies were obtained from those nodules. Pathologic examinations revealed "synovial sarcoma". As skeleton single photon emission tomography was unremarkable, primary pleuropulmonary synovial sarcoma was decided as diagnosis and chemotheraphy was planned for the patient. Primary pleuropulmonary synovial sarcoma is a rare neoplasm of lung and pleura but it is rare entity.

Key Words: *Pleuropulmonary sarcoma, thoracoscopy.*

Synovial sarcoma (malignant synovioma) is derived from the synovium or immature mesenchymal elements. This tumour accounts for 5-10% of all soft tissue sarcomas (1). It occurs mainly in the extremities and is closely related to tendons, tendon sheaths and bursal structures. It has also been described in locations unrelated to synovial tissues, including the head and neck, chest wall, pleura, lungs, heart, mediastinum, abdominal wall, kidneys, prostate and vagina.

Pulmonary sarcomas constitute only 0.1-0.5% of all primary lung malignancies. The most frequently reported subtypes of sarcomas in the lung are leiomyosarcoma, malignant fibrous histiocytoma, fibrosarcoma, and, more recently, synovial sarcoma (2). Pulmonary sarcomas may derive from the lung parenchyma, bronchial tree or pulmonary arteries. The most important entity in the differential diagnosis is metastatic synovial sarcoma. Because the morphologic features of primary and metastatic synovial sarcomas are similar, clinical and radiologic evaluation is essential to exclude the presence of a primary tumor outside the thorax.

Synovial sarcoma encompasses two histologic subtypes, monophasic and biphasic, with the monophasic variant being the more common. Nearly all synovial sarcomas have a specific t (X;18) (p11.2;q11.2) chromosomal abnormality (synovial sarcoma translocation, chromosome 18 [*SS18*]; alternative titles and symbols: synovial sarcoma translocated to X chromosome [*SSXT*]; *SYT*; *SYT-SSX1* fusion gene; *SYTSSX2* fusion gene) resulting in fusion of either of two variants of the *SSX* gene with the *SYT* gene. The genetic features might relate to morphology and outcome (3).

CASE REPORT

A 76-years-old woman was admitted for investigation of a low-grade fever, productive cough, dyspnea, weight loss and left-sided chest pain which had been present for one month. Her past medical history was unremarkable. She did not smoke cigarette or drink alcohol and had no family history of cancer. Physical examination revealed low-grade fever and absent breath sounds in the left lung field. The purified protein derivative skin test was negative. A chest computed tomography (CT) showed enlarged mediastinal lymph nodes, as well as a left-sided pleural effusion (Figure 1). Bronchoscopy showed only collapses in the left bronchial system without any endobronchial lesion. Transbronchial brushings were clear for microbiologic and cytologic examinations. Thoracentesis revealed hemorrhagic pleural effusion which was exudate and with lymphocyte predominancy. The blind

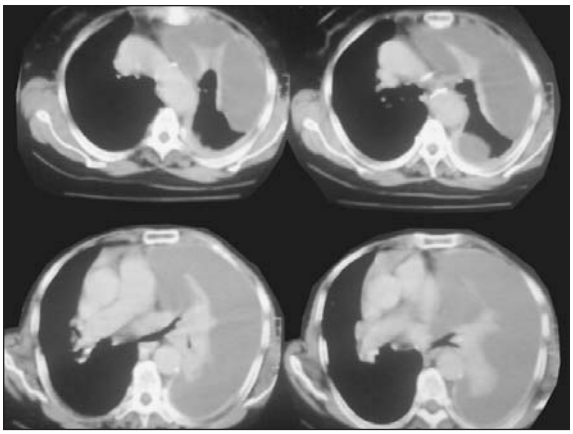

Figure 1. Chest CT showed enlarged mediastinal lymph nodes, as well as a left-sided pleural effusion.

parietal pleural biopsy via Abrams needle revealed chronic inflammation of pleura. During a left sided thoracoscopy performed under local anesthesia, total collapse of left lung and multiple pleural nodules were observed predominantly on posterior parietal pleura, biopsy samples were obtained from those nodules (Figure 2). At the same time talc pleurodesis was performed and thoracic tube dreinage was ended in second day after thoracoscopy. Pathologic examination of nodules was revealed "synovial sarcoma". Since the skeleton single photon emission CT performed to investigate any possible primary site was unremarkable, primary pleuropulmonary synovial sarcoma was accepted as diagnosis and chemotheraphy was planned for the patient.

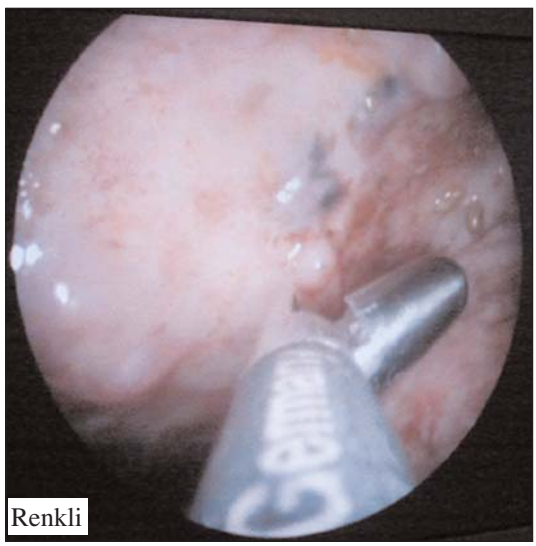

Figure 2. Left sided thoracoscopy performed under local anesthesia, total collapse of left lung and multiple pleural nodules were observed predominantly on posterior parietal pleura.

DISCUSSION

In the new edition of "Histological Typing of Soft Tissue Tumors" by the World Health Organization (WHO), synovial sarcoma is placed among the "miscellaneous soft tissue tumours" (4). Biphasic synovial sarcomas may be relatively easy to identify at light microscopy, but their differentiation from other biphasic tumors, including diffuse biphasic malignant mesothelioma, pleomorphic carcinoma of the lung, and pleuropulmonary blastoma, is essential. Solitary fibrous tumor, spindle cell carcinoma of the lung, diffuse sarcomatoid malignant mesothelioma, fibrosarcoma, leiomyosarcoma, cellular schwannoma, malignant peripheral nerve sheath tumor, and Ewing sarcoma are included into the differential diagnosis of monophasic synovial sarcoma. Separation of these entities is based on clinical history, radiologic distribution of disease, histologic features, histochemical and immunohistochemical staining, and cytogenetic and molecular studies (5,6). For our case, the macroscopic apperance in thoracoscopy was suggestive for a malignant pleural disease, but the final diagnosis was made by histhological features.

The radiologic manifestations of primary pulmonary sarcomas overlap with those of many other lesions of the lung and pleura, including primary and metastatic lung neoplasms, localized fibrous tumor of the pleura, malignant mesothelioma, and other rare primary parenchymal sarcomas (eg, malignant fibrous histiocytoma, fibrosarcoma, leiomyosarcoma, hemangiopericytoma, malignant nerve sheath tumors, and sarcomatoid carcinoma). Malignant mesothelioma usually is manifested as a circumferential pleural mass with or without associated pleural effusion. A history of asbestos exposure or the presence of contralateral pleural plaques is indicative of this diagnosis. Although the radiological findings of our case was not characteristic for malignant mesothelioma, left sided chest pain might be suggestive for that diagnosis even with the absence of the relevant expo-

sure history. At that point, histological discrimination is obligatory.

Factors predicting a worse prognosis for patients with primary pulmonary synovial sarcomas include older age (> 20 years), tumour size > 5 cm, the SYT-SSX1 variant, incomplete resection and a large number of mitotic figures (> 10 per 10 high-powered fields) (7). Our patient was an old woman, no resection could be performed on widely affected pleura.

There is no standardized therapy for patients with primary pulmonary synovial sarcoma; most patients are treated with extensive surgical resection, surgery and adjuvant radiation therapy or with surgery and adjuvant chemotherapy. Synovial sarcoma is chemosensitive to ifosfamide and doxorubicin, with an overall response rate of approximately 24% (8). The prognosis for patients with pulmonary synovial sarcoma is poor, with an overall 5-year survival rate of 50% (9).

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