Soft tissue sarcoma metastatic to pleura

Hüseyin YILDIRIM¹, Muzaffer METİNTAŞ¹, Güntülü AK¹, Emine DÜNDAR², Sinan ERGİNEL¹

¹ Osmangazi Üniversitesi Tıp Fakültesi, Göğüs Hastalıkları Anabilim Dalı,
² Osmangazi Üniversitesi Tıp Fakültesi, Patoloji Anabilim Dalı, Eskişehir.

ÖZET

Plevraya metastaz yapmış yumuşak doku sarkom olgusu

İnsan vücudundaki tüm kanserlerin plevraya metastaz yapabildikleri bilinmektedir. Bununla birlikte görülme sıklığı açısından erişkin kanserlerin %1’i’nden azını oluşturan yumuşak doku sarkomlarının plevra metastazları son derece nadir dır. Histopatolojik özellikleri temelinde yumuşak doku sarkomlarının sarkomatöz mezotelyomalardan ayrımının yapılması zordur. Burada ilerleyici nefes darlığı ve göğüs ağrısi yakınmaları ile kliniğimize başvuran ve yapılan torakoskopi biyopsisi sonrası yumuşak doku sarkomunun plevra metastazı tespit edilen 57 yaşındaki bir erkek hasta nadir görülen bir olgu olması nedeniyle sunulmuştur.

Anahtar Kelimeler: Yumuşak doku sarkomu, pleural metastaz, torakoskopi.

SUMMARY

Soft tissue sarcoma metastatic to pleura

Hüseyin YILDIRIM¹, Muzaffer METİNTAŞ¹, Güntülü AK¹, Emine DÜNDAR², Sinan ERGİNEL¹

¹ Department of Chest Diseases, Faculty of Medicine, Osmangazi University, Eskişehir, Turkey,
² Department of Pathology, Faculty of Medicine, Osmangazi University, Eskişehir, Turkey.

Almost all cancers can cause distant pleural metastases. However, pleural metastases of soft tissue sarcoma that constitute less than 1% of adult solid malignancy are extremely rare. It is very difficult to distinguish them from sarcomatous malignant mesothelioma on histopathological features. We report a 57 year-old man who presented to us with left chest pain and progressive dyspnea and was diagnosed to have a pleural metastases of soft tissue sarcoma by thoracoscopic biopsy.

Key Words: Soft tissue sarcoma, pleural metastases, thoracoscopy.

Yazışma Adresi (Address for Correspondence):
Dr. Hüseyin YILDIRIM, Osmangazi Üniversitesi Tip Fakültesi, Göğüs Hastalıkları Anabilim Dalı, Meşelik 26480 ESKİŞEHİR - TURKEY
e-mail: heyulu2002@yahoo.com
Metastases of malignant tumor spread to the pleura are common causes of pleural effusions. Currently, lung cancer is the most common metastatic tumor to the pleura in men and breast cancer in women (1). Soft tissue sarcomas (STS) are relatively uncommon cancers that constitute less than 1% of adult solid malignancy. STS can arise almost anywhere in the body. Distant metastases occur most often to the lung (2). There are infrequent reports of pleural effusion caused by sarcomas metastasizing to the pleura from an extra-thoracic primary. We present a case of a patient with soft tissue sarcoma who developed pleural metastases.

CASE REPORT

A 57-year-old man presented with a month history of left chest pain and progressive dyspnea. He had a 40-pack-year smoking history. His medical history was significant for STS (malignant peripheral nerve sheath tumor). Sarcoma had been diagnosed out of our institute in February 2005. We learned that the patient was treated with three cycles of chemotherapy (adriamycin, cisplatin, mitomycin) and radiotherapy. Subsequently, the disease recurred a year after chemo-radiotherapy, and he had undergone surgical amputation of the knee of the foot.

On hospital admission, a physical examination revealed dullness to percussion and reduced breath sounds on the left side and tachycardia with a regular rhythm. The patient was afebrile with a pulse rate of 102 beats/min and a BP of 140/90. The patient’s hemoglobin level was 10.9 g/dL, his total WBC count was 12,900/mm³, and his platelet count was 805,000/mm³. Serum electrolyte levels and renal function were normal. Serum lactate dehydrogenase level was elevated at 627 U/L. Relevant laboratory results were a significantly accelerated erythrocyte sedimentation rate (96 mm/h) and a slightly elevated C-reactive protein (11.5 mg/L). Arterial blood gas indexes included a PaO₂ of 71 mmHg, PaCO₂ of 39 mmHg, HCO₃⁻ of 27 mEq/L, pH of 7.36 and oxygen saturation of 94%.

The chest radiograph showed a massive left pleural effusion with a right mediastinal shift. A chest computerized tomography (CT) scan confirmed the presence of massive pleural effusion and diffuse pleural thickening on the left hemithorax (Figure 1).

A thoracentesis revealed thin, grossly hemorrhagic pleural fluid. The pleural fluid was lymphocytic exudates. Examination of the pleural fluid showed the following: pH 7.05; ADA 46 IU/L; lactate dehydrogenase level 2523 U/L; protein level 4.7 g/dL; and glucose level 18 mg/dL. No organisms were identified on Gram’s stain or culture, nor were malignant cells identified by cytology.

On day 3, medical thoracoscopy was performed to rapidly obtain an adequate specimen. Thoracoscopy revealed dense fibrous bands, and the nodular lesion was found to diffusely involve the parietal pleura. Biopsies were taken from the tumor on the parietal pleura (Figure 2). Over 2 L

Figure 1. Contrast-enhanced CT demonstrates the presence of massive pleural effusion and diffuse pleural thickening on the left hemithorax.

Figure 2. Thoracoscopic view showing dense fibrous bands and the nodular lesion in the parietal pleura.
of fluid were removed. After thoracoscopy, this fibrosis prevented the lung expanding completely, consequently 250,000 U of streptokinase in 100 mL normal saline solution during three day though the chest tube was instilled to destroy the adherences. Pleurodesis was not successful because complete apposition of pleural surfaces can not be achieved.

Biopsy specimen revealed a very dense population of spindle cells with fascicles intersecting at acute angles with nerve like whorls in some areas. It was also seen some hypocellular areas with wavy nuclei and frequent mitosis. Immunohistochemically, tumor cells were positive for vimentin, but negative for pancytokeratin, S100 and calretinin (Figure 3).

The patient general condition and functional capacity are not relevant for surgical treatment. Therefore, the pleural lesions were accepted medically unresectable. The patient then underwent adjuvant chemotherapy with a cycle of ifosfamide, adriablastin and mesna. Despite aggressive chemotherapy, his condition deteriorated further, and he died from progressive disease and respiratory failure 2 weeks after initiation of therapy.

**DISCUSSION**

Soft tissue sarcomas (STS) are rare malignant tumors that can occur in many parts of the body such as muscle, fat of the extremities or the trunk. There are many types of STS include fibrosarcoma, liposarcoma, malignant peripheral nerve sheath tumors, malignant fibrous histiocytoma, rhabdomyosarcoma and synovial cell sarcoma. They account for less than 1% of malignant neoplasm in general (3). Primary sarcomas of the lung and thorax can be rarely seen. Only 10% of patients with STS have distant metastases at presentation. Distant metastases usually occur within 2 to 3 years of initial diagnosis, and 40% to 60% of patients still develop metastatic disease after therapy. The lungs are a common site of metastases and are often the cause of death. Other potential sites of metastasis include bone, the brain, and the liver (2). However, effusions associated with pleural metastases are rare, especially so long after primary diagnosis. In their series involving 25 patients with malignant peripheral nerve sheath tumors, Kourea et al., reported a median survival of 8.5 month for these patients. Two of their patients had pleural metastases (4).
Metastases to pleura are more common than primary tumors of pleura. Malignant effusions result predominantly from obstruction and disruption of lymphatic channels by malignant cells (1). In most patients with usual STS, the dominant pattern of metastases is hematogenous. Lymph node metastases are rare; less than 5% show nodal spread (2). Hematogenous metastasis from STS is observed primarily in the lungs as randomly distributed nodular lesions. Consequently, it is generally accepted that pleural effusions do not develop when the pleura is involved by sarcomas because of the characteristic absence of lymphatic metastases (5).

The patient was accepted as metastases of STS to the pleura with the microscopic and immunohistochemical findings as well as the presence of history for this tumor. Histologically, STS may be difficult to differentiate from sarcomatous malignant mesothelioma. The distinction is especially important because of differences in management and prognosis. Since the neoplastic cells in sarcomatous mesothelioma consistently express cytokeratins, immunohistochemical staining is a useful method in differentiating STS from the sarcomatoid mesothelioma (6). In the presented case, tumor cells were positive for vimentin, but negative cytokeratin. This finding was sufficient to warrant the diagnosis of metastatic STS.

In general, treatment for STS depends on the stage of the cancer, and the patient’s age and general health. Soft tissue sarcomas are treated with multimodality therapy. Surgery continues to be the primary treatment method of STS. Surgical resection has now consistently been shown to prolong survival of patients with STS who develop pulmonary metastases (2). Radiotherapy is used to reduce local recurrence, especially for high-grade tumors or if the surgical margin was insufficient. The role of chemotherapy in the treatment of STS remains unclear. Recently, doxorubicin and ifosfamide are the most active single-agents in the therapy of sarcomas with response rate above 20%. Although recent advances in therapy have been encouraged, recurrences develop after curative local therapy (7).

Because of the rarity of these cases, there are no data concerning the treatment of pleural effusions caused by STS. However, we believe that treatment with drainage by a chest tube, with talc pleurodesis, will be very helpful to obtained symptomatic relief, especially when the patients general status are not suitable for surgical procedure.

In conclusion, we presented this case, because metastatic STS as the cause of a malignant pleural effusion are a rare occurrence.

REFERENCES