

Primary pleural leiomyosarcoma presenting as multiple pleural masses

Multipl plevral kitleler şeklinde kendini gösteren primer plevral leiomyosarkom

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ABSTRACT

In this article, we report a 74-year-old female patient with primary pleural leiomyosarcoma presenting with three pleural masses. Two diaphragmatic masses were completely excised via thoracoscopy. A positron emission tomography showed the apical mass as the primary site of origin, which was completely excised via axillary thoracotomy. As surgical margins were negative, patient was not given chemo- or radiotherapy, and is disease-free after 17 months of follow-up. Surgical resection is the mainstay of treatment of leiomyosarcoma. In nonmetastatic cases or when the surgical margins are negative, postoperative chemo- or radiotherapy may not be required. To our knowledge, there is no case of primary pleural leiomyosarcoma presenting with multiple pleural masses reported in the literature.

Keywords: Leiomyosarcoma; pleura; thoracotomy; video-assisted thoracic surgery.

Leiomyosarcomas are rare in the thoracic cavity. They can both affect the mediastinum and the lung. Pleural leiomyosarcomas (PLs) are unusual, and almost all of the cases are metastatic. The lesion is named as primary pleural leiomyosarcoma (PPL) when the pleura is the primary site. A limited number of individual cases with PPLs have been reported. Primary pleural leiomyosarcoma tends to form a pleural mass with or without pleural effusion. In this article, we report a case of PPL presenting with three separate foci in the parietal pleura.

ÖZ

Bu makalede, üç plevral kitle ile kendini gösteren primer plevral leiomyosarkomlu 74 yaşında bir kadın hasta sunuldu. İki diyafragmatik kitle torakoskopi ile tamamen eksize edildi. Pozitron emisyon tomografisinde primer köken alanın apikal kitle olduğu görüldü; bu kitle aksiller torakotomi ile tamamen eksize edildi. Cerrahi sınırlar negatif olduğundan, hastaya ameliyat sonrası kemo- veya radyoterapi verilmedi ve hasta 17 aylık takip sonucunda hastaliksiz durumdadır. Cerrahi rezeksiyon leiomyosarkom tedavisinin temelini oluşturur. Metastatik olmayan olgularda veya cerrahi sınırlar negatif olduğunda, ameliyat sonrası kemo- veya radyoterapi gerekli olmayabilir. Bilgimiz dahilinde, literatürde multipl plevral kitleler şeklinde kendini gösteren primer plevral leiomyosarkom olgusu bildirilmemiştir.

Anahtar sözcükler: Leiomyosarkom; plevra; torakotomi; video-yardımlı toraks cerrahisi.

CASE REPORT

A non-smoker 74-year-old female patient without a remarkable medical history presented with dyspnea and right-sided chest pain. She had a history of exposure to asbestosis since she was grown up in a rural area where the incidence of mesothelioma was high. Chest radiograph showed opacity on the right side. Computed tomography of the chest revealed an apical pleural mass (6.0x6.0 cm), two diaphragmatic pleural masses (3.0x3.0 cm and 2.0x1.0 cm), and right pleural



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Figure 1. Computed tomography. (a) An apical mass of 6.0x6.0 cm, a diaphragmatic mass of 3.0x3.0 cm with massive pleural effusion. (b) A third mass of 2.0x1.0 cm located on deep posterior part of diaphragm. Masses are shown with arrows.

effusion (Figure 1a, b). Thoracentesis demonstrated an exudative fluid. Nucleated cell count was 1000/mm³ (52% lymphocytes), total protein concentration was 45 g/dL (a pleural fluid-to-serum ratio of 0.63), lactate dehydrogenase level was 528 IU/L, and glucose level was 92 mg/dL. Cytological examination detected no tumoral cells, and the fluid culture results were negative. A video-assisted thoracoscopic surgery was planned for diagnosis.

The patient underwent general anesthesia and double-lumen endotracheal intubation, and was placed in left lateral decubitus position. Two separate trocar incisions were used. Two liters of blood-stained fluid was removed. The two abovementioned

diaphragmatic masses were completely excised. Due to apical adhesions, no attempts were made on the apical lesion. She had an uneventful course, and was discharged on the second postoperative day. Histological examination exhibited spindle cell proliferation in cords and sheets, nuclear polymorphism, and infrequent mitosis (Figure 2a). Immunohistochemically, the tumor was stained positive for alpha-smooth muscle actin, desmin, vimentin, and S-100 protein, and negative for calretinin, carcinoembryonic antigen, cytokeratin, leukocyte common antigen, neuroendocrine filament, and CD-117 (Figure 2b). Therefore, both lesions were diagnosed as pleural leiomyosarcomas. To exclude a

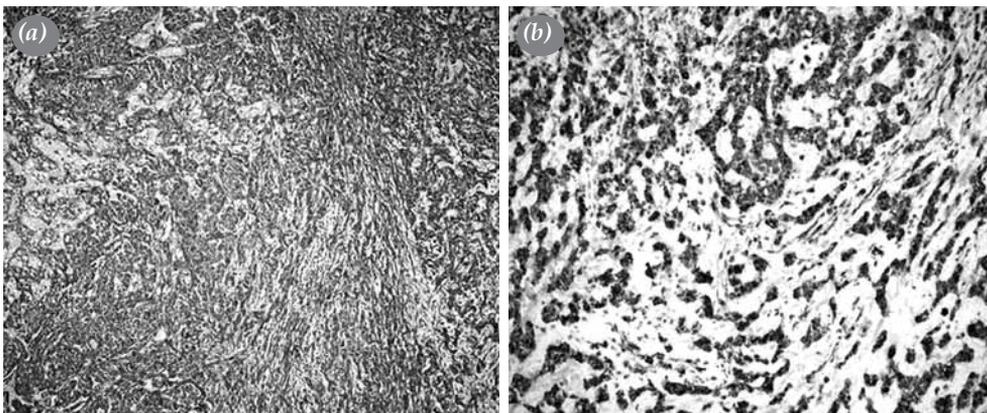


Figure 2. Pathological findings of resected tumor. (a) Neoplastic spindle cell proliferation arranged in interlacing bundles and fascicles, as well as infrequent mitosis (H-E x 10). (b) Immunohistochemical staining demonstrating positive reaction with actin (SMA x 20).



Figure 3. Positron emission tomography-computed tomography demonstrating apical mass lesion as the only pathologically metabolically active lesion in body with a maximum standardized uptake value of 9.3.

metastatic disease, a positron emission tomography was performed. The maximum standardized uptake value of the remaining apical lesion was 9.3, and since there was no other possible tumor focus, the final diagnosis was a PPL (Figure 3). The patient underwent an axillary thoracotomy after one month, and the apical lesion was totally excised. The pathological examination was consistent with the previous ones, with clear surgical margins. The patient was discharged three days after the surgery. Since the surgical margins were negative, and the patient was old, postoperative chemotherapy and/or radiotherapy were not given. The patient is alive and without recurrence after a follow-up of 17 months.

DISCUSSION

Leiomyosarcomas are cancers of smooth muscle cells, mostly arising from the uterus, gastrointestinal tract, or soft tissue. Thoracic leiomyosarcomas are rare, and commonly seen in the mediastinum, heart, and lung.^[1] Pleural leiomyosarcomas are unusual, and fewer than 10 cases have been reported.^[2] Most PLs are metastatic from uterus, gastrointestinal tract, and retroperitoneum.^[3] It is named PPL if the pleura is the primary site.^[2-5] The pathological examination of diaphragmatic masses in our case demonstrated a PL. The positron emission tomography demonstrated no other primary focus than the apical lesion, thus the final diagnosis was PPL.

Pleural leiomyosarcoma behaves like other primary tumors of the pleura in symptomatology

(dyspnea, chest pain, and cough), physical signs, and radiology.^[4,5] The tumor very rarely encases the lung like mesothelioma, but mostly forms a mass with or without pleural effusion.^[2-5] Our case had three separate masses in the parietal pleura, and to the best of our knowledge, this is the first reported case of PPL with multiple foci in the English literature. It is known that leiomyosarcomas mostly spread to the body via hematogenous metastasis.^[1] Since the parietal pleura is very rich in vascular network, it is possible for a PPL to spread along the pleura using this network. We think that the larger apical lesion was the primary focus in our case, and the two smaller diaphragmatic lesions were the pleural metastases.

Histological examination with immunohistochemical staining is required to differentiate PL from other more frequent pleural malignancies, such as malignant sarcomatoid mesothelioma, malignant fibrous histiocytoma, pleural solitary fibrous tumor, and neurogenic tumors.^[2] Microscopically, leiomyosarcomas are characterized by malignant spindle cells with scant fibrillary cytoplasm arranged in chords and sheets with a variable mitotic activity. Leiomyosarcomas are nearly uniformly positive for smooth muscle actin, desmin, and vimentin, and negative for calretinin, carcinoembryologic antigen, cytokeratin, leukocyte common antigen, neuroendocrine filament, and CD-117.^[3,4] The histological and immunohistochemical characteristics of the lesions in this case demonstrated a PL.

There is no optimal treatment for PL. The advised treatment is surgical resection alone or with adjuvant radiation or chemotherapy. The need for adjuvant radiotherapy or chemotherapy depends on the tumor grade and the clinical stage of the patient.^[3-5] Chemotherapy is indicated in case of a locally advanced or metastatic disease.^[1] The most important factor affecting overall survival is margin status.^[2,5] We performed surgical resection in this case, since the disease was not metastatic, and the complete surgical resection seemed technically possible. The resected margins were tumor-negative. Thus, the patient was not given any additional chemo- or radiotherapy.

It is known that PPL is extremely rare. To our knowledge, no such case with multiple pleural foci has been presented in the literature. When a complete surgical resection with tumor free margins is achieved, there may be no need for postoperative chemo- or radiotherapy in elderly patients.

Declaration of conflicting interests

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