



Primary sclerosing epithelioid fibrosarcoma of the pleura

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TO THE EDITOR:

Sclerosing epithelioid fibrosarcoma (SEF) is a rare variant of fibrosarcoma that occurs primarily in the deep musculature and is often associated with the adjacent fascia or periosteum.⁽¹⁻⁴⁾ We searched the Medline, LILACS, SciELO, and MD Consult databases for articles on SEF published in the last 20 years.

Here, we report the case of a previously healthy 31-year-old female patient who presented with dyspnea in 2012. She was a former smoker with a smoking history of 14 pack-years. She sought emergency room treatment. Physical examination revealed decreased breath sounds at the left lung base and a Karnofsky performance status of 80%.

The thoracic surgery team requested a chest CT scan, which revealed a lung mass and a pleural lesion. Pleural and lung biopsies were therefore performed. Pathological findings were suggestive of sarcoma or sarcomatoid mesothelioma. Further investigation included CT scans of the chest, abdomen, and pelvis, as well as magnetic resonance imaging of the chest. Chest CT and magnetic resonance imaging findings included extensive pleural effusion with anterior septation; contrast-enhanced tissue formations in the parietal pleura, the largest of which was 8.5 cm × 2.0 cm and located posterior to the basal segments on the left; left lung volume loss; and intraparenchymal nodules scattered throughout the left lung parenchyma, the largest of which was 1.7 cm.

A decision was made to perform a pneumonectomy with thoracotomy and mediastinal lymphadenectomy. Intraoperative findings included visceral and parietal pleural involvement (affecting the underlying left lung), as well as contiguous invasion of the lower third of the anterior chest wall (ribs 7 to 10), left diaphragm, and pericardium. A decision was made to perform a pleuropneumonectomy with en bloc resection of part of the left diaphragm, pericardium, and mediastinal lymph nodes (stations 5, 6, 7, 8L, and 9L), as well as thoracotomy with resection of the affected ribs, the resection margins being macroscopically free of disease (Figure 1). The left main bronchus stump was stapled with a 75-mm stapler, and the chest wall was reconstructed with polypropylene mesh (Prolene®; Ethicon, Somerville, NJ, USA). Closed pleural drainage was performed. The patient stayed in the ICU for 6 days, being discharged on postoperative day 7. Pathological findings were consistent with locally advanced SEF of the pleura. The affected lymph nodes (5 of 12 lymph nodes) were resected, the resection margins being free of disease. Histopathological examination revealed a neoplasm composed of ovoid epithelioid cells

with hyperchromatic nuclei and inconspicuous nucleoli embedded in a densely collagenized stroma (Figure 2). Immunohistochemistry was negative for S-100 protein, CD34, calretinin, Wilms tumor 1 oncogene product, and desmin but positive for mucin-4, findings that are consistent with primary SEF of the pleura. Over the course of a 3-year follow-up period, CT scans of the chest were performed every 6 months, the last follow-up CT examination having shown mild left pleural effusion and no signs of local recurrence.

SEF was first described by Meis-Kindblom et al. in 1995⁽⁵⁾ as a rare, slow-growing, deep soft tissue sarcoma that is composed of epithelial tumor cells arranged in nests embedded in a hyalinized fibrous stroma and that affects individuals in the 14- to 87-year age bracket, affecting both genders equally.⁽¹⁾ It is primarily located in the lower extremities, pelvic and pectoral girdles (39%), and trunk (21%).⁽¹⁾ One third of cases present as painful, deep-seated intramuscular masses associated with the adjacent fascia or periosteum.⁽³⁾ Although SEF is histologically classified as a low-grade sarcoma, it is clinically aggressive. Local recurrence and metastases are observed in 30-50% of cases; however, systemic spread is usually delayed for 5 years or more.⁽⁴⁾ The most common metastases (in 43-86% of cases) are to lung, bone, chest wall/pleura, pericardium, brain, scalp, breast, and liver.^(4,5) With regard to 36-month prognosis, 34% die from SEF, 35% remain alive with disease, and 31% remain alive without disease. It appears that the primary site of disease is associated with prognosis, the 36-month mortality rate being 46% in patients with SEF of the head and neck, 38% in those with SEF of the upper extremities, and 26% in those with SEF of the trunk.⁽¹⁾ In addition, it appears that neither gender nor tumor size influence the prognosis of SEF.⁽²⁾ We found no case reports of primary SEF of the pleura.

Ossendorf et al.⁽¹⁾ reported that distant disease was independent of tumor size; however, no patient with a primary tumor of 5 cm or less initially presented with metastases. Given the lack of randomized studies, there is no consensus regarding the optimal treatment for SEF.⁽¹⁻⁴⁾

The major histological features of SEF include nests and cords of rounded epithelioid cells surrounded by hyalinized collagenous stroma. The cells are relatively uniform, with scanty cytoplasm; the nuclei are eccentric and pleomorphic, being ovoid, elongated, or angulated.⁽¹⁾

The differential diagnosis includes hyalinizing spindle cell tumor with giant rosettes, low-grade fibromyxoid sarcoma, sclerosing lymphoma, synovial sarcoma, fibroma, and fibrosarcoma.^(1,3) Local recurrence and distant

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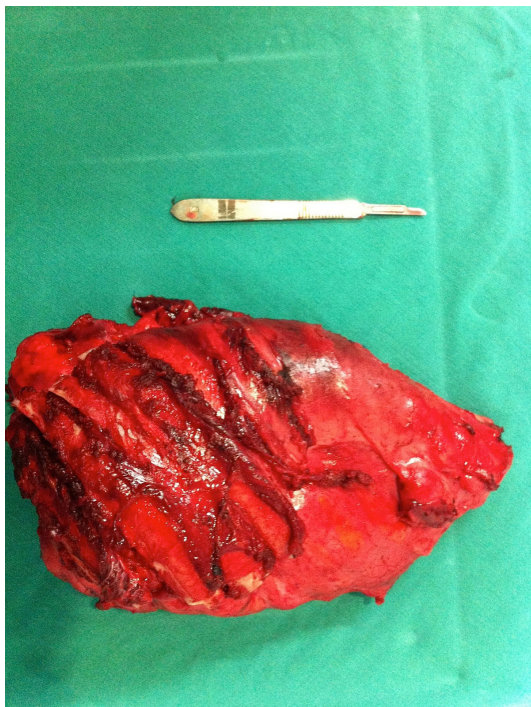


Figure 1. Surgical specimen showing en bloc resection of the left lung, pleura, and ribs.

metastases (most commonly to the lung, pleura, and bone) are observed in > 50% and > 40% of cases of fibrosarcoma, respectively.⁽⁵⁾

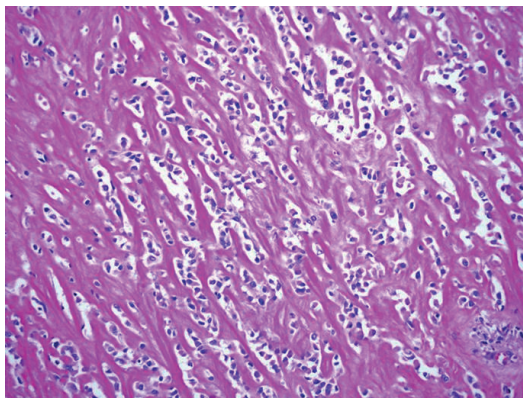


Figure 2. Photomicrograph showing a neoplasm composed of ovoid epithelioid cells with hyperchromatic nuclei and inconspicuous nucleoli embedded in a densely collagenized stroma (H&E; magnification, ×40).

An extremely rare tumor that few pathologists have encountered, SEF is often difficult to diagnose. Despite being low grade, SEF is a clinicopathologically distinct tumor with malignant potential, the rates of recurrence, metastasis, and mortality being considerable.^(1,4)

As is the case with soft tissue fibrosarcomas, surgery is the treatment of choice because other treatment options have little efficacy. Further studies are needed in order to determine whether the clinical progression of primary SEF of the pleura is the same as that of other SEFs, as well as to determine the best form of treatment for patients with primary SEF of the pleura.

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