Case Report

Primary synovial sarcoma of pleura: A case report and review of the literature

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ABSTRACT

Primary pleural synovial sarcomas (SSs) are rare tumors comprising <1% of all primary lung tumors. They are frequently misdiagnosed as relatively more common entities such as malignant mesothelioma. We present one such rare case of biphasic SS in the pleura of a 40-year-old female who presented with massive right hemothorax, and computed tomography revealed a pleural-based tumor in the right lung. Histopathological examination of the excised tumor was suggestive of biphasic SS. Immunohistochemical staining showed positivity with antibodies against cytokeratin 7, epithelial membrane antigen, vimentin, and bcl-2.

Key words: Immunohistochemical, pleura, primary, synovial sarcoma

INTRODUCTION

Synovial sarcoma (SS) is an uncommon soft tissue tumor that occurs primarily in the extremities of young adults, especially in the periarticular region. However, it is also reported to occur in the head and neck, mediastinum, heart, esophagus, lungs, abdominal wall, mesentery, and retroperitoneum.[1-4] SS of the pleura more often represents metastatic disease from a primary soft tissue tumor.[3] Primary SS of the pleura is rare[1-4] and comprises <1% of all primary lung malignancies.[5] They are frequently misdiagnosed as relatively more common entities such as malignant mesothelioma (MM).

CASE REPORT

A 40-year-old female patient presented with chief complaint of chest pain and shortness of breath after a history of fall 7 days back. She had no respiratory complaints previously. On palpation, tenderness in 5th and 6th rib area was present. Air entry was absent in the right side of thorax on auscultation. Chest X-ray was suggestive of massive right hemothorax. Intercoastal drain was inserted and 2.5 L of clotted blood was drained. Contrast-enhanced computed tomography thorax and abdomen revealed heterogeneously enhancing lobulated soft tissue mass along mediastinal, diaphragmatic, and right costal pleura more so along diaphragmatic pleura indenting the superior border of liver [Figure 1]. Right side loculated hydropneumothorax with mediastinal lymphadenopathy was also seen. Possibility of pleural metastasis from an unknown primary was suggested. The patient underwent thoracotomy for evacuation of clots, and soft tissue mass was excised. Histopathological examination of the excised masses revealed sweeping and intersecting fascicles of uniform spindled cells with ovoid to elongated nucleus with tapering ends, pale cytoplasm with indistinct cell borders. At places, these spindled cells were separated by ropy collagen bundles. In addition, variable-sized glandular spaces lined by flattened to cuboidal epithelium and containing homogenous granular/eosinophilic, periodic acid-Schiff (PAS) positive diastase-resistant secretions in the lumen were identified [Figures 2 and 3]. There were frequent mitotic figures (>10/10

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high power field), extensive areas of tumor necrosis, focal hemangiopericytomatous areas, mast cells, and hyalinized blood vessels. Immunohistochemical (IHC) analysis revealed positivity for cytokeratin (CK) 7, epithelial membrane antigen [Figure 4], vimentin [Figure 5], and bcl-2 [Figure 6]. Staining for calretinin was negative.

DISCUSSION

SS occurs primarily in the extremities of young adults, especially in the periarticular region. However, they can also occur in the head and neck, mediastinum, heart, esophagus, lungs, abdominal wall, mesentery, and retroperitoneum.[1-4] SS of the pleura is more commonly due to the metastatic disease from a primary soft tissue tumor.[3] Primary SS of the pleura is rare[1-4] and comprise < 1% of all primary lung malignancies.[5] Extensive literature review revealed that the patients with primary pleural SS ranged in age from 9 to 69 years and 57% were male. Chest pain was the most frequent complaint followed by dyspnea, cough, pleural effusion, and pneumothorax. Radiologically, a mass and/or pleural effusion was present in most cases. Macroscopically, the tumors ranged from 4.5 to 21 cm in size and were mostly solid and localized pleural-based masses. Areas of necrosis, hemorrhage, calcification, and cystic change were commonly described. A total of 50% of the cases were biphasic tumors.[6] Distinction of biphasic SS from biphasic MM can be extremely challenging, but is very important as these entities have distinct treatments and prognosis. SS typically form localized, pleural-based masses with or without pedicles. In rare cases of SS, multifocal patterns have also been reported.[7] MM, in contrast, typically present as multiple nodules covering the pleura or as a diffuse sheet-like pleural thickening that can encase and compress the lungs.[8] Histologically, biphasic SS and biphasic MM exhibit subtle differences.[4] Biphasic SS have a long interweaving spindled component that is compact and cellular, with little stromal collagen. Foci of hemiangiopericytomatous architecture and of microcalcification are characteristic, and hyaline fibrosis

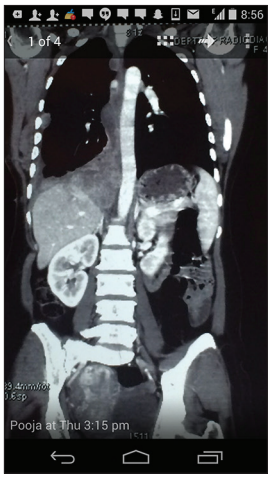

Figure 1: Contrast-enhanced computed tomography thorax and abdomen revealed heterogeneously enhancing soft tissue mass along mediastinal, right costal, and diaphragmatic pleura indenting the superior border of liver

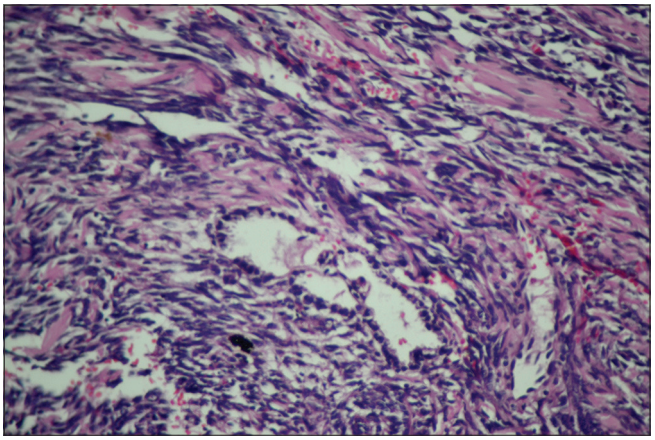

Figure 2: Photomicrograph showing spindle cell as well as epithelial components (H and E, ×40)

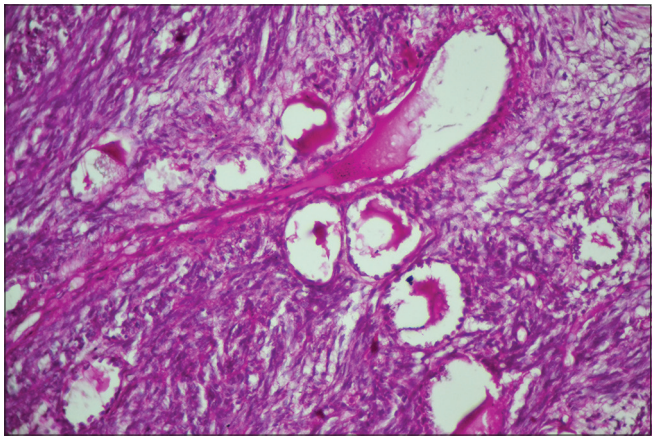

Figure 3: Periodic acid-Schiff-positive diastase-resistant secretions in the glandular lumina (periodic acid-Schiff, ×40)

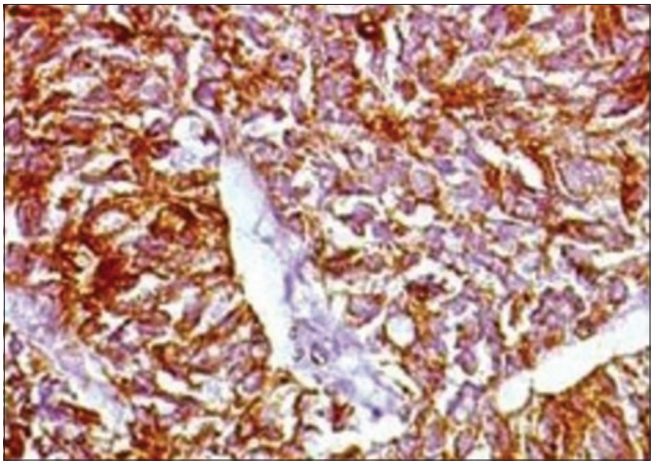

Figure 4: Epithelial membrane antigen positivity in tumor cells (immunohistochemical, ×40)

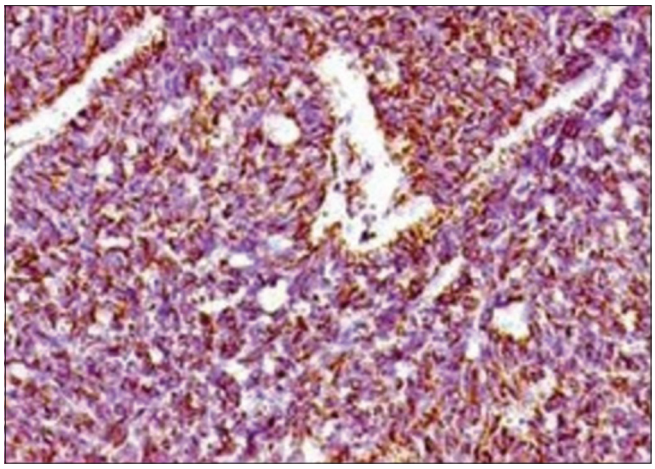

Figure 5: Vimentin positivity in tumor cells (immunohistochemical, ×40)

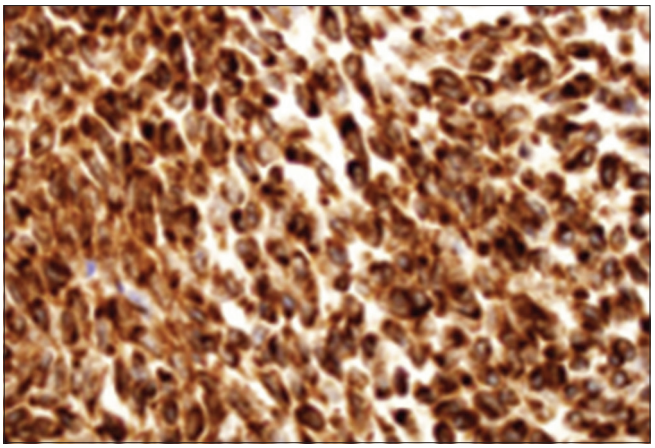

Figure 6: Bcl-2 positivity in tumor cells (immunohistochemical, ×40)

can be present. Mast cells are often prominent, but glycogen is sparse. In contrast, the spindled component of biphasic MM consists of shorter, looser fascicles of blunt spindle cells with more stromal collagen. Hyaline fibrosis and hemangiopericytomatous architecture are rare. Mast cells are also fewer in number, but glycogen is abundant. The epithelial component of biphasic SS typically consists of epithelial cells forming cleft-like glandular spaces and tubulopapillary structures. The epithelial component of well-differentiated biphasic mesotheliomas can also be tubulopapillary, but there is typically a gradual transition between the sarcomatous and epithelial elements in these tumors, while there is a sharp abutment of these areas in SS.[8] In contrast to MM, pleural SSs contain secretions that are mucicarmine-positive and hyaluronidase-resistant and PAS-positive and diastase-resistant.[4] Rare MM may exhibit mucicarmine or PAS staining, but it is eliminated with hyaluronidase or diastase digestion. Overall, the use of IHC in the distinction between MM and SS of the pleura is challenging and limited at best. Panels of markers are recommended since no single marker is diagnostic of either MM or SS. However, co-expression of bcl-2, CD56, and CD99 with negative staining for calretinin, WT-1, and focal CK positivity strongly suggests the diagnosis of SS rather than MM. On electron microscopy, biphasic and epithelioid MMs are characterized by long, slender, tortuous branching microvilli, but this finding may be diminished or lost in poorly differentiated neoplasms. Abundant intracytoplasmic glycogen is also seen. SS, in contrast, have shorter blunt microvilli and glycogen is sparse to absent. In those cases with overlapping clinical, gross, histologic, and IHC features, the diagnosis can be established by cytogenetic studies as the chromosomal translocation t (X;18) (p11.2;q11.2) is characteristic of SS. The best treatment for SS of the pleura remains unclear. Because a SS of the pleura is rare, the paucity of data regarding its natural history and the limited number of published cases, its treatment is similar to other localized SS's treatment. A multidisciplinary approach, including surgery, chemotherapy, and radiotherapy has been suggested.[9] Surgery (radical resection) is the first treatment of SS. Adjuvant radiotherapy is usually recommended after incomplete resection or after extensive resection of large tumors.[9,10] The benefits of chemotherapy are unclear; however, improvement in survival has been described with doxorubicin and ifosfamide.[11] Neoadjuvant chemotherapy could be an appropriate option to allow the radical surgical procedure by reducing tumor volume and potentially treating micrometastasis disease.[11] Radiotherapy, chemotherapy, and radiofrequency thermal ablation may be considered as alternative treatments for inoperable patients.[12] Other options have been reported, Abe *et al*.[10] suggest combined hyperthermia therapy with chemoradiotherapy for the patient with advanced inoperable primary pleural SS. SS is an aggressive tumor with a very poor prognosis compared to other sarcomas. Recurrences are common and patients may require multiple surgical resections. A 5-year disease-free period of 20.9%[13] has been reported.

CONCLUSION

SS of pleura is a rare tumor and frequently mistaken for MM which is the most common malignant tumor arising from pleura. As a result, diagnosis is easily overlooked since these entities have overlapping clinical, gross, histologic, and IHC features. In addition, there is no gold standard therapy because SS is a rare malignancy and not enough data are available due to the few cases that have been quoted in literature.

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Conflicts of interest

There are no conflicts of interest.

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