

Primary Pleural Leiomyosarcoma A Common Tumor in Uncommon Site - A Case Report

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Abstract

Leiomyosarcomas (LMS), the malignant neoplasm showing smooth muscle differentiation, are aggressive and can arise from various locations. It commonly arises from uterus, gastrointestinal tract and soft tissue. Primary Pleural Leiomyosarcomas cases have been reported rarely. Articles suggest that it presumably originates from bronchial or pulmonary vascular smooth muscle and exclusion of all other possible primary sites of leiomyosarcoma is important before signing out the pathological report. Because of its rarity there is a limited knowledge about the natural history and prognosis. This is a rare case report of leiomyosarcoma presenting as primary in pleura.

Date of Submission: 05-04-2022

Date of Acceptance: 20-04-2022

I. Case presentation

This is a case of a 70 year old gentleman, an ex – smoker and by profession a carpenter presented to our hospital with a history of cough and dyspnoea for 3 months. There wasn't any history of fever or hemoptysis. He is a known case of CAD. No other relevant past history obtained. His breathe sounds were decreased on right side and blood investigations within normal limits. His chest x-ray (Figure 1) shows an opaque right hemithorax and CT thorax shows an enhanced mass lesion involving right hemithorax abutting the posterior chest wall suggestive of solitary fibrous tumor of pleura and right upper lobe collapsed due to mass effect. Whole body PET CT showed a large heterogeneously enhancing pleural based soft tissue mass involving the right posterior hemithorax based on right posterior costal/posterior mediastinal pleura. No definite scan evidence of abnormal hypermetabolism/ lesion elsewhere in the body.

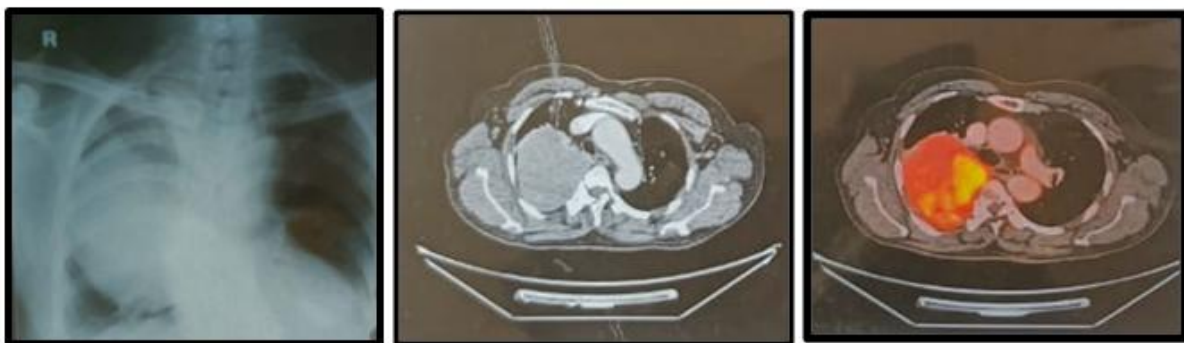


Figure 1: CHEST XRAY, CT THORAX, PET CT

Excision of the mass was done. Intraoperative finding was a large mass measuring 12 x 10 cm arising from the pleura adherent to the lung. We received a lobulated firm to hard grey brown mass measuring 13 x 11 x 8 cm. Cut section showed a grey white lobulated mass with necrotic areas (Figure 2). Gross differentials considered were solitary fibrous tumor (SFT) and mesothelioma.

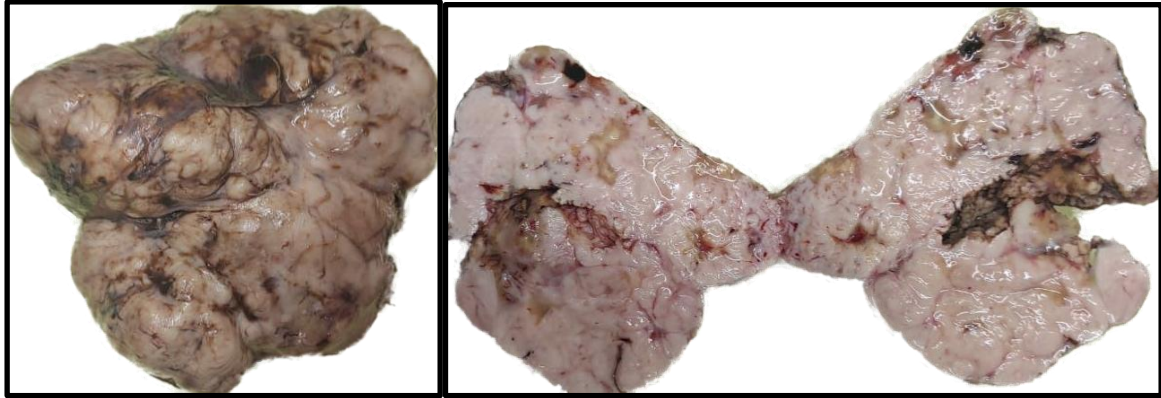


Figure 2

Histopathology showed (Figure 3) a tumor arranged as interlacing fascicles composed of pleomorphic spindle cells with atypical hyperchromatic nucleus ,(Figure 4) atypical mitosis and tumor cell necrosis were present.

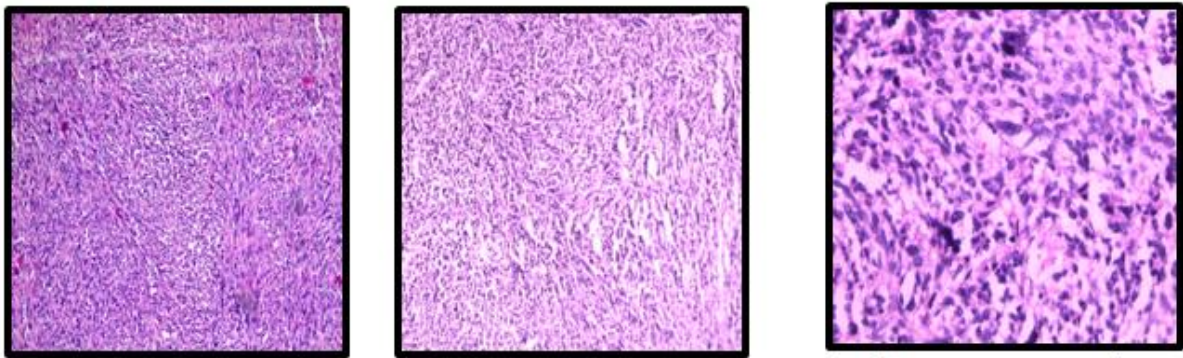


Figure 3 : 4x, 10x, 20x

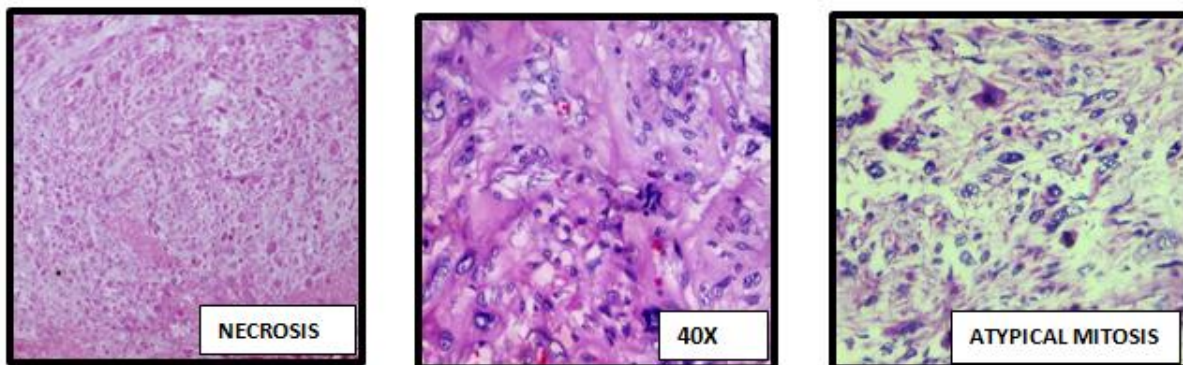


Figure 4

Immunohistochemistry was done. The tumor cells were positive for SMA and negative for calretinin and CD34 (Figure 5) ,ruling out differentials like malignant sarcomatous mesothelioma , (SFT) ,synovial sarcoma which are more common and confirms the diagnosis of leiomyosarcoma. Since we could exclude any other lesions in the body, the diagnosis was given as primary pleural leiomyosarcoma.

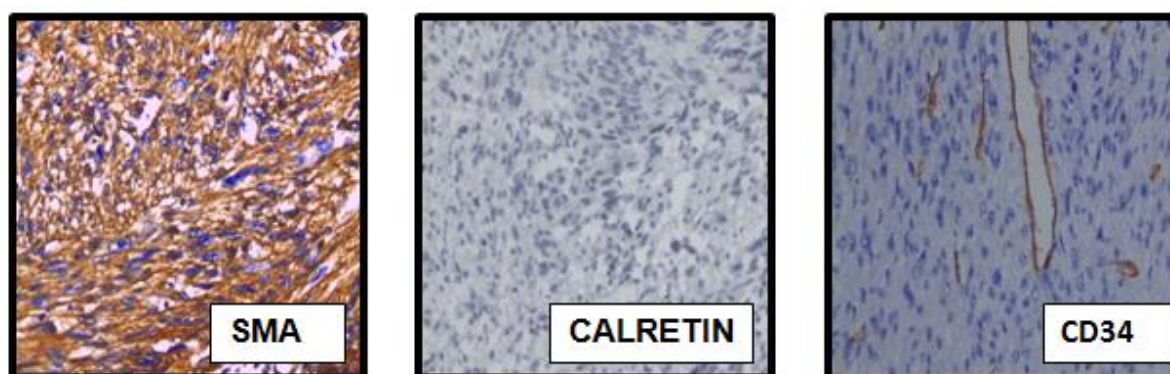


Figure 5

II. Discussion

Leiomyosarcoma originating from the pleura is a rare entity. LMS would originate either from the smooth muscle cells or from the mesenchymal stem cells that differentiate into smooth muscle cells. It is mandatory to rule out leiomyosarcoma in other parts of the body before concluding the report as primary. IHC slides have been found useful as supplementary to the histological slides in diagnosis of LMS as well as to exclude other primary malignancies of pleura as mentioned earlier.

Surgical removal of the mass is the recommended main stay of therapy. As per the literature, histological grade and clinical stage are the best prognostic factors and the median survival for patients with extensive disease is deduced to be 12 months. The decision on adjuvant chemoradiation can be determined on the base of grade and stage of the tumor. Usual chemotherapy drugs comprise trabectedine, doxorubicin and ifosfamide, and the overall response rate is approximately 20%.

This case report is intended to point out the importance of considering the rare entities when deriving differentials for a pleural mass.

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Milu Elsa Paul, et. al. "Primary Pleural Leiomyosarcoma A Common Tumor in Uncommon Site - A Case Report." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 21(04), 2022, pp. 01-03