Primary pleural leiomyosarcoma - A rare entity

Sir,

Leiomyosarcomas, are aggressive malignant tumors of the smooth muscle cells and can arise from any location, but commonly take origin from uterus, retroperitoneum, or other intra-abdominal viscera. Primary pleural leiomyosarcomas are uncommon, and only a few cases have been described till now. It is imperative to establish the primary site of origin of the leiomyosarcoma, to provide appropriate therapy. Before establishing the definitive diagnosis of pleural leiomyosarcoma, all primary sites of origin have to be excluded. If there are no signs of systemic metastasis, pleural resection surgery has been proposed for tumor management.[1]

A 40 year old non smoker, previously healthy female, presented with sub-acute, progressive shortness of breath, dry cough and left sided pleuritic chest pain for the past one and a half months. General examination revealed an average built female with insignificant findings. Chest auscultation revealed absent breath sounds in left infra-mammary, infra-axillary and infra-scapular areas. Chest skiagram showed left sided massive effusion. Routine blood tests were normal. The patient underwent removal of around 1200 ml of pleural fluid at a peripheral hospital, which revealed a lymphocytic, exudative effusion with mesothelial cells and a few atypical cells. Contrast enhanced computed tomography (CECT) of the chest revealed a massive left sided pleural effusion with nodular thickness, contralateral mediastinal shift and no mediastinal lymphadenopathy. The patient was referred to our center for further diagnostic procedures, and underwent medical thoracoscopy with pleural biopsy. Gross examination of the pleura revealed multiple fleshy, lobulated pleural nodules of varying sizes with no visible nodules on the lung, along with hemorrhagic effusion [Figure 1]. Multiple pleural biopsies were taken. The cytospin examination of pleural fluid revealed a few spindle shaped atypical cells. Fibreoptic bronchoscopy did not reveal any mass lesion or mucosal abnormality. The patient underwent a FDG-PET/CT for disease staging which revealed, extensive homogenously enhancing lobulated nodular thickening with increased FDG uptake (SUVmax-12.3) involving the left costal, mediastinal and ipsilateral diaphragmatic pleura [Figure 1]. There was passive collapse of underlying lung parenchyma and mild mediastinal shift towards the contra-lateral side. Calcified mediastinal, paratracheal, subcarinal and left hilar lymph nodes with no significant FDG uptake were seen. Histopathology of the Pleural biopsies revealed a malignant tumor of spindle to oval cells with no features of differentiation and few tumor cells showing prominent nucleoli suggestive of malignant spindle cell tumor. Immunohistochemistry (IHC) staining was done with cytokeratin 5/6, 7, and 20, smooth muscle actin, vimentin, calretinin, CD-117, TTF1, CD-34, and S-100 protein to further characterize the exact nature of the tumor. The tumor cells stained strongly positive for smooth muscle actin and vimentin and negative for all other markers [Figure 2], ruling out other differentials like sarcomatous mesothelioma (calretinin), solitary fibrous tumor (CD-34), and neurogenic sarcoma (S-100 protein) and confirming the diagnosis of malignant leiomyosarcoma.

Post thoracoscopy and ICD tube placement, pleurodesis with slurry was done and ICD was removed after 3 days. The patient's condition was discussed in the tumor board and planned local radiation and Doxorubicin based chemotherapy. Surgical resection was ruled out by the thoracic surgeon owing to the extensive pleural involvement. The patient was given first cycle of doxorubicin based chemotherapy, after which the attendants refused further definitive therapy.

The, clinical and radiological features of these tumors are similar to various other pleural neoplasms and are difficult to diagnose accurately even on standard light microscopy.[2] Most of these tumors are diagnosed by thoracoscopic or thoracotomy aided biopsies.[3] Histologically, leiomyosarcomas are characterized by malignant spindle cells with scant fibrillary cytoplasm

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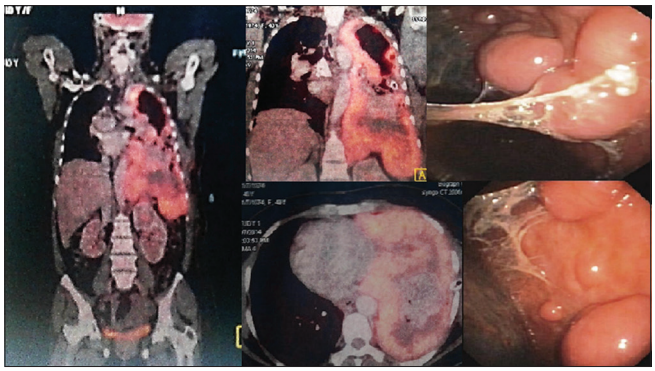

Figure 1: PET CT demonstrating FDG uptake in the pleura, corresponding thoracoscopic pictures of the pleural masses

and cigar-shaped nuclei with variable mitotic activity. The closest differential is the sarcomatous mesothelioma and to accurately diagnose the leiomyosarcoma the use of an IHC antibody panel is recommended. These tumors are positive for smooth muscle actin, desmin, and vimentin and negative for carcinoembryonic antigen, cytokeratin, calretinin, leukocyte common antigen, neuroendocrine filament, CD-117 and S-100 protein.[3]

As the optimum therapy of these tumors is not known, the exact treatment should be decided by a tumor board. Surgery has been tried in patients with limited disease. Multimodality therapy with surgery, radiation and chemotherapy has been described in literature.[1] Chemotherapy is offered to patients with high mitosis, disseminated disease and those who cannot be offered surgery owing to medical or surgical inoperability.[4] Usual chemotherapy regimens comprise trabectedine, doxorubicin and ifosfamide, and the overall response rate is approximately 20%.[4,5] Poor prognosis is usually predicted by advanced clinical stage and a high mitosis on the biopsy, indicating a high grade malignancy.[4]

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Conflicts of interest

There are no conflicts of interest.

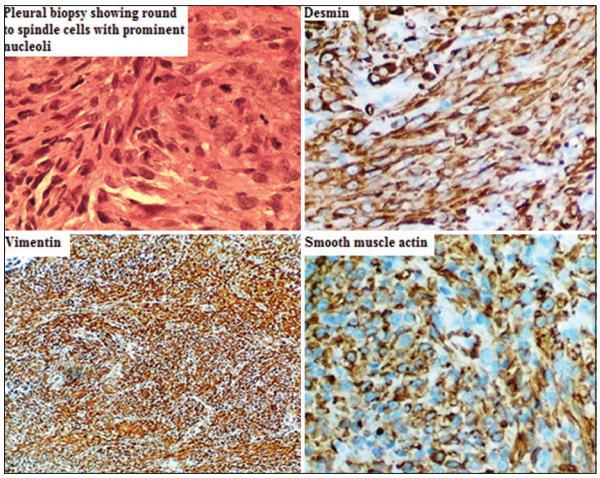

Figure 2: Histopathology and IHC panel, showing positivity for vimentin and smooth muscle actin

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