Pleomorphic sarcoma of the diaphragm-A rare case

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ABSTRACT- We present the case of a 72 year old male with a left sided intrathoraccic mass, which was reported on histopathology as a pleomorphic sarcoma of the diaphragm- a rare tumour at this site.

Key words- Diaphragm, Intrathoracic mass, Pleomorphic sarcoma

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INTRODUCTION

We present a case of a 72 year male, chronic smoker, with a history of breathlessness, left sided chest pain, cough, loss of appetite and weight loss for one month. Radiological findings showed a large left pleural effusion and on CT scan there was a large benign mass lesion in the left lower zone. The mass was excised and reported as a high grade pleomorphic sarcoma of the diaphragm. Diaphragmatic tumours are rare [1,2] with isolated reports of diaphragmatic malignant fibrous histiocytoma [3,4] which was the original name for pleomorphic sarcoma.

CASE REPORT

A 72 year male, chronic smoker and hypertensive patient presented to the Surgery OPD with a history of breathlessness, left sided chest pain, cough, loss of appetite and weight loss since one month. Vital signs were normal. Respiratory examination showed decreased air entry on left side. Cardiovascular and abdominal examinations were normal. Routine blood investigations were normal. Chest X ray showed massive left pleural effusion with shift of mediastinum to right side. Pleural fluid examination showed predominantly lymphocytes with reactive mesothelial cells. No malignant cells were seen. CT scan chest showed a large well defined multi-cystic mass with smooth margins, in the left lower zone of the thoracic cage measuring 15x13x10 cm. Bilateral lung parenchyma was normal. There was eversion of left dome of diaphragm with displacement of spleen and left kidney anteriorly. Large left pleural effusion was

noted. There was no lymphadenopathy. Visualized skeletal system was normal. The impression on CT scan was a large benign mass lesion in the left lower zone

On bronchoscopy, right bronchus and right lung were normal. There was external compression of left upper and left lower bronchus. Mucosa was normal. A bronchio alveolar lavage was done which was negative for malignancy. A thoracoscopic biopsy was taken from the left chest mass. Biopsy revealed spindle cell proliferation singly and in short haphazard fascicles, arranged in a collagenous stroma. Occasional cells showed atypia and few mitotic figures were seen. Heamangiopericytomatous vascular pattern was noted. There was no evidence of lung parenchyma. It was suggestive of a benign spindle cell tumour. Differential diagnosis given was solitary fibrous tumour and inflammatory myofibroblastic tumour.

Patient underwent surgical excision of mass through left postero-lateral thoracotomy. Intra-operatively it showed a huge mass arising from left dome of diaphragm. It was free from lung and pleura. The tumour was removed piecemeal. The specimen was sent for histopathological examination. Gross examination showed multiple bulky pieces of soft tissue total measuring 20 x16 x14 cm and weighing 2.5kg. Histopathologically the tumour was composed of spindle shaped cells with oval nuclei and scanty cytoplasm. Nuclei showed hyperchromatism and pleomorphism. Many multi-nucleate giant cells were seen (Figure 1 and 2).

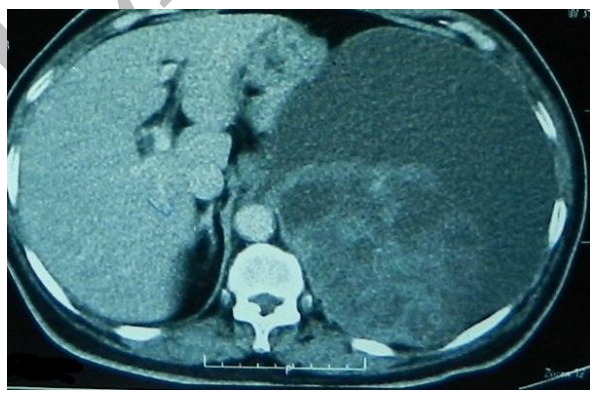

Figure 1: CT thorax showing a large well defined multi-cystic mass with smooth margins in the left lower zone of the thoracic cage measuring 15x13x10 cm

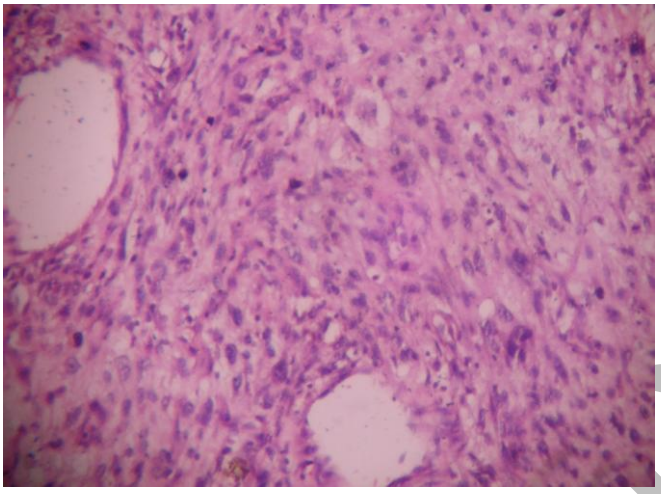

Figure 2: Photomicrograph showing oval to spindle shaped cells with nuclear pleomorphism and multi nucleate giant cells. [Hematoxylin and Eosin stain, X400]

At places cells were arranged in fascicular patter. Myxomatous areas, small areas of calcification and few large blood vessels surrounded by tumour cell were seen. There were also areas of necrosis, haemorrhage and increased mitotic activity. The histopathology was suggestive of a pleomorphic spindle cell sarcoma. Immunohistochemistry (IHC) was done which was positive for EMA and SMA (weak) and negative for Desmin, CD 34, S-100 protein, Calretinin, WT-1and Cytokeratin. Hence based on IHC the final diagnosis was High Grade Pleomorphic Sarcoma.

The patient was advised chemotherapy which he refused and went home. On further follow up we were informed that he expired 4 months later.

DISCUSSION

In 1868, Grancher reported the first case of primary diaphragmatic tumour as an autopsy finding and diagnosed it as fibroma. Wiener and Chou in 1965 reported the first case of a tumour of the diaphragm. [1] Another study reviewed 37cases of MFH in the chest wall, which had previously been reported in the Japanese and English literature [2]. There are isolated case reports of diaphragmatic MFH. [3, 4]

Malignant fibrous histiocytoma was the terminology previously used for undifferentiated high grade pleomorphic sarcoma [UPS]. [5] It was a diagnosis of exclusion and for many years the most common soft tissue tumour in adults. With the advance in technology and knowledge,

the original MFH has been sub classified into various differentiations like, leiomyosarcoma, rhabdomyosarcoma, liposaroma and now this term pleomorphic malignant fibrous histiocytoma is reserved for a small group of undifferentiated pleomorphic sarcomas. As a consequence, the apparent incidence of pleomorphic MFH has fallen sharply over the past 10 years. Clarification is important because it allows patients to receive specific treatments such as trabecidin in myxoid/round cell liposarcoma. [6]

In 1992, Fletcher published a retrospective series of 159 cases diagnosed as pleomorphic sarcoma from the files of the Histopathology Department, St. Thomas's Hospital, and London. These tumors were re-assessed morphologically, using meticulous tumour sampling, immunohistochemistry, and ultra-structurally. [6] Only 13% of the cases in this retrospective series were truly eligible to be diagnosed as MFH.

Sarcomas arise from mesenchymal stem/progenitor cells and account for 1% of all malignancies. Human mesenchymal stem cells are the progenitors of undifferentiated pleomorphic sarcoma. MFH comprises a heterogeneous group of poorly differentiated neoplasms. At that time two possible origins of UPS were considered likely: (i) that it arises from a primitive pluripotential mesenchymal cell that demonstrates different extents of differentiation; (ii) that it is a nonspecific entity of poorly differentiated neoplasms of different types.

Most undifferentiated high grade pleomorphic sarcomas occur in the extremities (especially the lower limb) and less often the trunk. The majority of cases arise in deep (subfascial) soft tissue, while less than 10% are primarily subcutaneous. A notable exception among pleomorphic sarcomas is dedifferentiated liposarcoma which is most common in the retroperitoneum. [5] The diaphragm is a rare site as seen in or case, and has been reported only in isolated cases [3, 4]

MFH has a high propensity for local recurrences (44%) and distant metastasis (42%).The pleomorphic sarcomas metastasize via blood and lymphatics relatively infrequently and towards the late stage of the disease. The commonest site is the lung, with an incidence of 30%, and the distant lymph node metastases occur in about 15% of cases. With diaphragmatic tumors, metastasis is virtually the rule, and lung and node metastases are relatively common.[7]

The prognostic factors that are known to adversely impact survival in patients with MFH include a tumor size > 5 cm, deep-seated location, high histological grade, and a high stage, based on the American Joint Committee on Cancer Staging System. Going by these criteria our patient had a poor prognosis, and hence expired 4 months after the surgery after refusing chemotherapy.

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