MESENCHYMAL TUMORS OF LUNG - A RETROSPECTIVE CLINICOPATHOLOGIC STUDY

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Abstract:
Mesenchymal tumors comprise a rare group of pulmonary tumors, constituting less than 5 percent of lung tumors. It includes various benign, malignant and hamartomatous lesions. A retrospective study was conducted in our department and all the pulmonary tumors on records were analyzed for a period of 6 years from January 2007 to December 2012. The various details analyzed were the presenting complaints, age, sex, radiological findings, gross morphology and microscopic findings. A total of nine cases belonged to this category and they included inflammatory myofibroblastic tumor, leiomyosarcoma, malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma sclerosing hemangioma.

Keyword:
lung - mesenchymal tumors - inflammatory myofibroblastic tumor - sclerosing hemangioma

INTRODUCTION:
Variety of benign and malignant tumors arise in the lung. Majorities are carcinomas and only about 5% of them are of mesenchymal origin. Mesenchymal tumors of the lung include tumors of fibrohistiocytic origin, tumors of muscular origin, hamartomatous tumors, osteochondromas, lipomatous, neural, and vascular tumors. It also includes miscellaneous tumors like sclerosing hemangioma. The incidence for these tumors is very low and primary pulmonary sarcomas account for less than 1 % of all primary lung tumors [1].

MATERIALS AND METHODS:
A retrospective study was conducted in our department and all the pulmonary tumors on record were analyzed for a period of 6 years from January 2007 to December 2012. The various details analyzed were the presenting complaints, age, sex, radiological findings, gross morphology and microscopic findings. The specimens included both excision specimens and tru-cut biopsies. The
specimens were fixed in 10 % neutral buffered formalin and embedded in paraffin. A 5 µ tissue sections were made and the slides were stained with Hematoxylin & Eosin routinely.

RESULTS:
The lung mesenchymal tumors in our study, included 2 cases of inflammatory pseudotumour (22%), 1 case of undifferentiated pleomorphic sarcoma (MFH) (11%), 1 case of leiomyosarcoma (11%), 3 cases of malignant peripheral nerve sheath tumor (34%) and 2 cases of sclerosing hemangioma (22%) – as depicted in table 1. A male predominance was observed as shown in table 2. Most of them were tru-cut biopsies, with few being lobectomy specimens. Among the two cases of inflammatory myofibroblastic tumor (Fig.1, 2), age incidence varied from 2nd to 6th decade. One case of MFH (Fig.3) occurred in a 60 years old male and it was positive for vimentin (Fig. 4), negative for S100 (Fig.5) and SMA. One case of Leiomyosarcoma (Fig.6, 7) was reported in a 37 years old male, which showed positivity for vimentin, desmin & SMA. 3 cases of MPNST (Fig. 8, 9) reported in males of 2nd and 3rd decade and were positive for S100 (Fig.10). The 2 cases of sclerosing hemangioma (Fig.11, 12) showed an age incidence of 3rd to 5th decade.

Table 1: Incidence of mesenchymal lung tumours
Table 2: Sex ratio of mesenchymal lung tumors
Inflammatory Myofibroblastic Tumor (Fig.1,2)
Fig. 1 Fig. 2
Malignant Fibrous Histiocytoma (Fig. 3, 4, 5)

Fig. 3 Fig. 4 – MFH - Vimentin Positivity
DISCUSSION:
INFLAMMATORY MYOFIBROBLASTIC TUMOUR:
It is also known as inflammatory myofibroblastic tumor, plasma cell granuloma, fibrous histiocytoma, fibroxanthoma and pseudosarcomatous myofibroblastic tumor. Most patients are under the age of 40 and many are children [2]. Half of the patients are asymptomatic, but some present with cough, hemoptysis, chest pain or shortness of breath. Chest x-ray shows a solitary.
peripherally located, sharply circumscribed mass without true encapsulation. Grossly those with more fibroblasts are firmer and gray-white, those with an increased number of plasma cells are tan and rubbery and those with abundant fat-filled macrophages have a more brilliant yellow-orange color and are soft and more friable. They are pulmonary lesions that are histologically similar to localized areas of organizing pneumonia. Characteristically, these lesions show filling of air spaces by plump fibroblastic cells, foamy histiocytes and chronic inflammatory cells such as lymphocytes and plasma cells.

**MALIGNANT FIBROUS HISTIOCYTOMA:**
They present in the sixth or seventh decade of life and show no sex predilection [2]. About one third of patients have cough, chest pain, and hemoptysis or weight loss. The others are asymptomatic. Chest radiographs show single or multiple nodular masses. Grossly, these tumors tend to be large. On cut surface, they are lobular, tan to white and can have some cavitated areas. Microscopically, MFH is characterized histologically by heterogeneous cell populations that include histiocytes, fibroblasts, pleomorphic giant cells and undifferentiated mesenchymal cells arranged in variable degree of storiform pattern. Necrosis is frequent and lymphocytes are found scattered. Mitoses are usually numerous and often atypical.

**LEIOMYOSARCOMA:**
Most patients are middle-aged to elderly and the tumors are round, well circumscribed, intraparenchymal fleshy masses that can be up to 24 cm in diameter and may invade the chest wall [2]. Their color varies from gray to tan. Necrosis and hemorrhage may be seen grossly in larger tumors. Cystic change may be present. Microscopically intersecting bundles of spindle-shaped cells occasionally arranged in storiform, palisaded or hemangiopericytoma-like patterns can be seen. Areas of hyalinization, hypcellularity and coagulative necrosis are frequent in larger tumors. The nuclei are characteristically elongated and blunted-ended. Nuclear hyperchromatism may be pronounced. Mitoses can usually be found.

**MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR (MPNST):**
The MPNST is typically a disease of adult life, most occurring in patients of 20–50 years of age [3]. Patients with NF1 develop these tumors at an earlier age. It is a rare and aggressive sarcoma that arises from the nerve sheath or shows features of nerve sheath differentiation. It arises as a large fusiform or eccentric mass in a major nerve. It is usually large more than 5 cm in diameter and has a fleshy, opaque, white-tan surface with areas of hemorrhage and necrosis. They have markedly irregular contours. Microscopically they show serpentine shape of the tumor cells; arrangement in palisades or whorls; with marked contrast between the deeply hyperchromatic nuclei and the pale cytoplasm (‘punched out nuclei’); perivascular concentration of tumor cells with a plumper shape; epithelioid appearance of the endothelial cells of these vessels; presence of large gaping vascular spaces, resulting in a hemangiopericytoma-like appearance; and geographic areas of necrosis with tumor palisading at the edges. Mitoses are usually abundant. S-100 protein, the most widely used antigen for neural differentiation, can be identified in 50–90% of MPNST [3].
SCLEROSING HEMANGIOMA:
It predominantly affects young adult women and typically appears as a well-defined, round, intraparenchymal soft-tissue mass in a juxtapleural location [4]. Most patients with sclerosing hemangiomas are asymptomatic. When symptomatic, patients usually present with vague chest pain and hemoptysis. Chest radiographs show nodularity. Grossly, they are well circumscribed and easily shell out from the adjacent lung parenchyma. They vary in color depending on the amount of fresh and old blood within them. Microscopically, these tumors are quite variegated, but basically consist of two types of cells: dark surface cuboidal cells and deeper round to polygonal paler cells. Architecturally, they are cellular solid zones, or papillary, or both solid cellular and sclerotic. Other common features include calcification and intratumoral haemorrhage [4]. There may be cholesterol clefts with foreign body type of giant cells around them.

CONCLUSION:
Mesenchymal tumors of lung are uncommon and include a wide variety of benign and malignant tumors. CT helps determine the location and features of the lesions and depicts associated findings to help document the extent of disease[5]. Proper histopathological interpretation of them is essential for assessing their biological behavior and for treatment.

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