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Solitary Fibrous Tumour: A rare site

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ABSTRACT
Solitary fibrous tumor (SFT) is a rare tumor of mesenchymal origin that account for less than 2% of all soft tissue masses[(1)]. Initially identified in the pleura, SFT has been identified in multiple anatomic locations and can arise anywhere in the body[(1)]. However, an intrapulmonary site of this kind of tumors is even rarer.[(2)] These tumors are often an incidental finding at standard chest X-ray. The definitive diagnosis is made after histopathological evaluation and the surgery is the best way to obtain simultaneous diagnosis and treatment especially that the fine-needle aspiration biopsy or bronchoscopic biopsy.[(2)]In our paper, we presented a case of a 24 year old male with generalized left sided chest pain and pleural effusion. CECT chest revealed an intrapulmonary mass lesion which on histopathological examination was confirmed to be a Solitary fibrous tumour (Hemangiopericytoma).

I. INTRODUCTION

Hemangiopericytoma (HPC) was described in 1942 by Stout and Murray as a distinctive soft tissue neoplasm, presumably of pericytic origin, exhibiting a characteristic well-developed 'staghorn' branching vascular pattern.[(5)] Over the years, it appeared that this growth pattern was a non-specific one, shared by numerous, unrelated benign and malignant lesions, and that HPC was better considered as a diagnosis of exclusion. Solitary fibrous tumor represents a single spectrum of mesenchymal tumors, of which hemangiopericytoma is a now considered a cellular phenotypic variant. [(3)]

Because of the major histological overlap between SFT and HPC, and lack of clear criteria to determine if a lesion should be called SFT or HPC, pathologists have been gradually abandoning the term 'hemangiopericytoma' in favor of the term SFT, so that most lesions that were called hemangiopericytoma 15 years ago tend to be called SFT now[(5)(12)]

SFT tumors may be found in almost any site of the body with intra-thoracic being the most common location followed by intra-abdominal. Within the thoracic cavity the majority are pleural-based followed by lung parenchymal tumors, then the mediastinum and the diaphragm. They arise equally in the right and left chest.[(1)]

II. CASE REPORT

A 24 year old male came to the Department of Respiratory medicine , Hamidia Hospital with the chief complain of left sided chest pain for 2months, dull and diffuse in nature, cough since 1 month, dry in nature., difficulty in breathing since 15 days.

On physical examination left sided pleural effusion was suspected. USG chest reported gross effusion of 5 cm. 3 litres of hemorrhagic fluid was drained.

CECT chest reported left lower lobe mass of size 4x3x2 cm . On fibreoptic bronchoscopy , shining globular mass was confirmed in the left lower lobe obliterating the opening of its anterior and lateral segments.

He underwent lobectomy. Specimen was sent to the Department of Pathology for histopathological evaluation.

The Department of Pathology, Gandhi Medical College, received a greyish brown to greyish black globular mass. It was 4x3x1.5cm in measurement. On cut section, it was cystic with wall thickness varying from paper thin to 0.3mm.

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Fig.1.2:Gross: A globular, grey brown to grey black mass(Department of Pathology, Gandhi Medical College, Hamidia Hospital, Bhopal)

Multiple sections were passed. On microscopic examination,there were areas of hypocellularity and hypercellularity separated by the "staghorn" like vessels. The lumina of the prominent gaping staghorn vessels were filled with variable number of red blood cells. The cells were bland spindle shaped with with central oval nuclei having blunt tips with small inconspicuous nucleolus and
diffuse chromatin. These were arranged perpendicular to the long axis of the blood vessels. Some areas of necrosis and myxoid degeneration between the cells were
also appreciated.

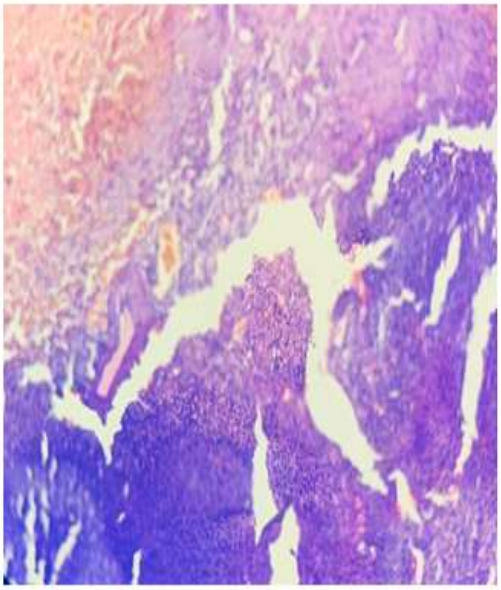

Fig.1.3:**10x: Hypocellular and hypercellular areas separated by "staghorn" like vessels with their lumina filled with red blood cells(Department Of Pathology, GMC, Hamidia Hospital,Bhopal)**

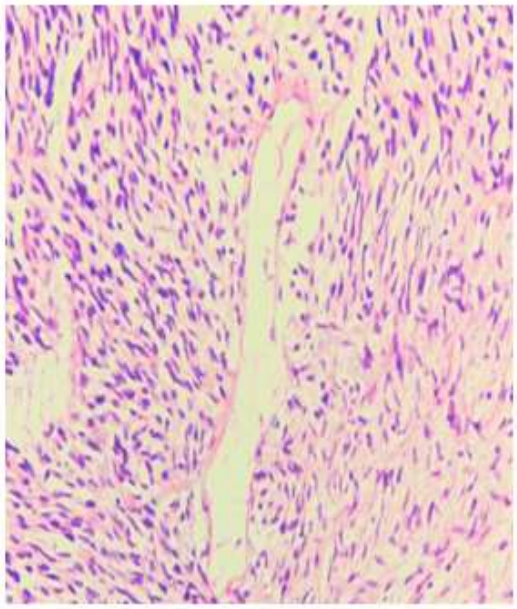

Fig.1.4: 40x:Bland spindle shaped cells with nuclei.arranged perpendicular to the long axis of the vessel(Department of Pathology, GMC, Hamidia Hospital, Bhopal)

A diagnosis of Hemangiopericytoma with cystic degeneration (Solitary fibrous tumour) was given.

III. DISCUSSION

Hemangiopericytoma (HPC) is a rare tumor originating from Zimmerman capillary

pericytes surrounding the endothelium.[7] Hence, HPC may develop anywhere with endothelium. Pericytes are contractile cells present normally, localized around the capillary and the post capillary venulae, It is most commonly seen in the head and neck. This is followed by the lower extremity and the retro peritoneum. [10]The most common extra pleural site and second most common location overall is the abdomen[1]. In our case, it was intra-pulmonary in location.

The clinical features depend on the site, size, and malignant potential of the tumor.[13] Common symptoms include dyspnea, cough, and a thoracic distress in the lungs and urinary retention in the pelvis or in the retroperitoneal site.[8][9] According to the site involved, epistaxis, proptosis, and nasal or sinus congestion may be observed[10]. Rarely, SFT may present with paraneoplastic syndromes, the most common being non-islet cell hypoglycemia. This is because the tumor produces high molecular weight insulin-like growth factor (IGF), specifically IGF-II. Seventy percent of these tumors will exhibit malignant behavior and presence of non-islet cell hypoglycemia is an overall poor prognostic indicator.[10]. Our patient had generalized dull chest pain alongwith dry cough and breathlessness. There were no other symptoms.

These tumors are often found incidentally at standard chest X-ray.[13] The definitive diagnosis is made after histological evaluation and the surgery is the mainstay to obtain simultaneous diagnosis and treatment .

Advances specifically in immunohistochemistry and molecular diagnostics have identified CD34 antibody, which is an endothelial marker as the most consistently expressed one in SFT, however even this lacks specificity for exact identification. More recently the discovery of the NAB2STAT6 fusion gene has led to more precise diagnosis of SFT[1]. Prognosis of SFTs also depend on morphologic and pathologic findings.[12] Benign and pedicled tumors have the best prognosis than malignant and sessile tumors[2]. In our case, it was a pedunculated tumour as observed on CECT chest and showed no malignant features on HPE.

Surgical management has been the mainstay treatment for SFTs. Obtaining adequate negative margins decreases the rate of recurrence and improves survival, alongwith a reduction in progression to metastasis.[6] . There have also been multiple retrospective studies evaluating the effectiveness of standard cytotoxic chemotherapy with doxorubicin-based regimens.[10]

IV. CONCLUSION

In conclusion, hemangiopericytoma is better to be considered as a diagnosis of exclusion because most hemangiopericytoma-like tumors are best regarded as variants of solitary fibrous tumor. In conformity with the new designation, our case should be included in the cellular variant of solitary fibrous tumor. The outcome and prognosis of patients diagnosed with such a tumor has a wide range of variability depending on many factors, which are still under debate; future studies are due to bring to light the pathologic correlations between the histologic features and the clinical course in individual patients.

Regardless of the name assigned to the lung tumor, patients with hemangiopericytoma/solitary fibrous tumor necessitate a multidisciplinary approach to their care, that requires involvement of nutritional and social services, behavioural medicine, and some home or hospital care as appropriate. A better general understanding of this disease process is expected to meet the needs of patients with malignant forms of hemangiopericytoma/solitary fibrous tumor.

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