

Primary Pulmonary Hemangiopericytoma

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Abstract

Case Report

Primary Pulmonary (lung) Hemangiopericytoma is a very rare neoplasm. It is known to be malignant tumor [1]. Hemangiopericytoma is known to be more malignant when the disease site is lung compared to soft tissues. The tumor is almost asymptomatic but in advanced stage it becomes symptomatic. Symptoms are generally similar to other cancers of lung like shortness of breathlessness, dyspnea, cough, fever. Tumor is usually treated with surgical resection. Prognosis is usually controversial.

Keywords: Primary Pulmonary Hemangiopericytoma, Very Rare Neoplasm.

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INTRODUCTION

Hemangiopericytoma is a rare mesenchymal neoplasm [1]. It develops from pericytes present in capillary walls [2]. They can be found in any part which have capillaries [3]. Initially were considered to be tumors of superficial structures (subcutaneous tissues and limb muscles) later discovered to be found in internal structures.³ It develops mainly in subcutaneous structure, skin, muscles of extremities [1]. It is rare in lungs, trachea, and mediastinum [1].

Primary growth within lung is rare and if present it is with malignant potential [3]. This mostly affects older population with asymptomatic presentation and non calcified solitary mass on X ray [4] usually size < 5cm in diameter [3]. Few patients have vascular disease related symptoms [5].

There are no established guidelines for their diagnosis and management [5]. Malignant disease is recognized by increased tumor size, increased mitotic rate, necrosis and hemorrhage [4]. Its diagnosis is based on histopathological examination⁵ since other methods such as CT and MRI have not proven to be accurate.

CASE REPORT

A 64 year old female presented to Radiation Oncology OPD at GMCH with chief complains of Chest Pain and Shortness of breath for 1 month. Pain was insidious in onset, moderate in nature, radiating to right arm, no aggravating or relieving factors were present

Patient also presented with shortness of breath, which was insidious in onset, initially associated with sternous activity, progressive, now associated with normal daily activities.

Patient also presented with fever which was associated with chills and rigor. Fever was undocumented with no diurnal variation.

Investigations

X-Ray

- Ill defined opacities are seen in right lung fields.
- Left CP angle is blunted suggestive of effusion.



Fig 1: X ray showing Left CP angle blunt

USG Whole Abdomen

- Liver- parenchyma shows fatty changes in both lobes.
- Gall Bladder- not visualize
- Pancreas- normal in size and echopattern.
- Aorta\ IVC appear normal.
- No ascites, no retroperitoneal lymphadenopathy
- Spleen- normal
- Right Kidney- Normal
- Left kidney- normal

CECT

- Contrast enhanced study shows large wedge shaped roundish mass 6.9* 6.5* 5.7 cm seen in right upper lobe.
- It is seen involving apical and anterior segment reaching upto anterior chest wall laterally.
- Medially extending upto right hilum.
- Laterally lesion is seen causing mild diffuse pleural thickening especially anterior however adjoining bones and chest wall appear normal.
- Another nodule 10*10 mm is seen in right lower lobe apico-posterior segment abutting posterior chest wall.
- Few small subcentimetric nodes seen with heterogenous attenuation in aorto- pulmonary window measuring~10mm.
- Note is made of enlarged lymph node in supraclavicular region on left side



Fig 2: Shows CECT findings

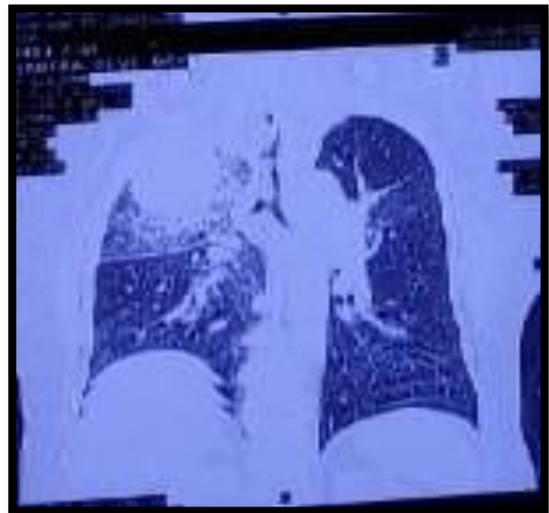


Fig 3: Shows CECT findings

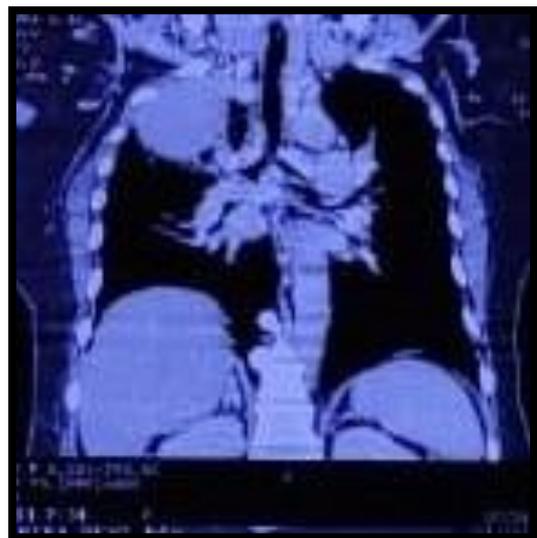


Fig 4: Shows CECT findings

Histopathological examination

Right Lung, Upper Lobe mass:- Hemangiopericyoma

- An invasive tumor arranged haphazardly or in ill defined fascicles.
- Tumor cells are spindle shaped having elongated nuclei with bland nuclear chromatin and scant to moderate amount of cytoplasm with indistinct cell borders.
- Dilated, branching staghorn like blood vessels with arrangement of tumor cells around them.

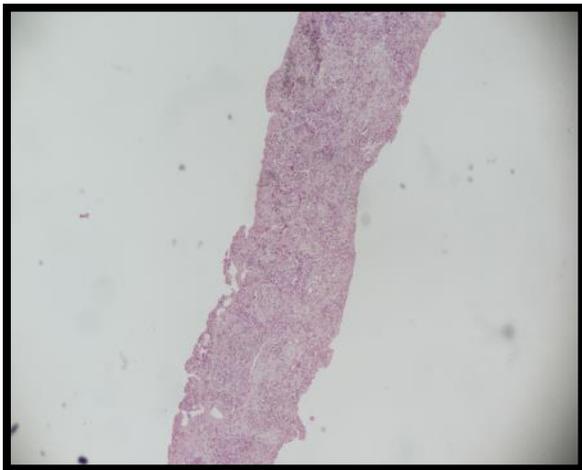


Fig 5: HPC 40x

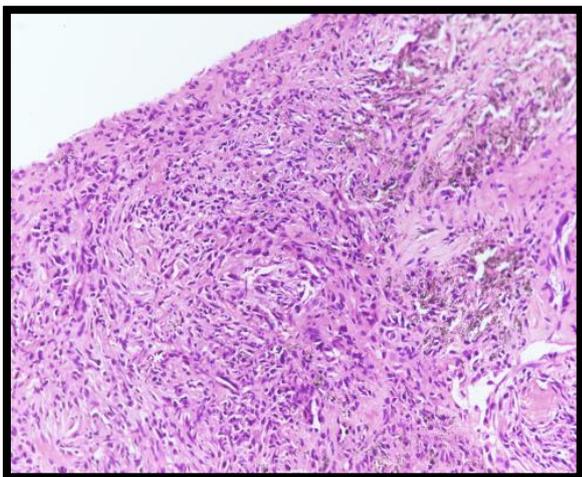


Fig 6: HPC 200x

IHC is positive for

1. CD34

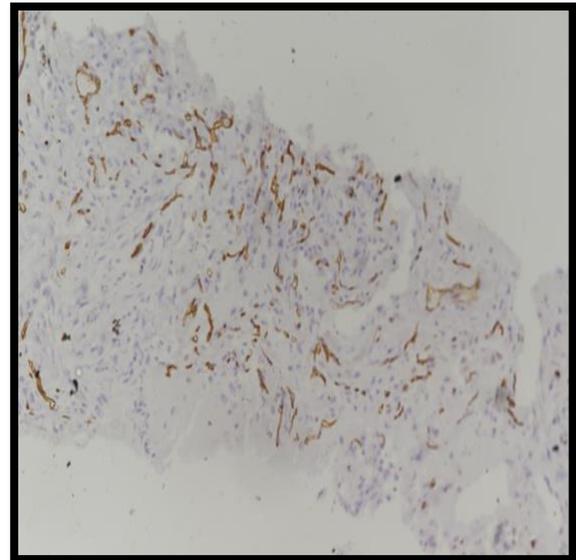


Fig 7: Picture showing staghorn vessels

Negative for

1. S100

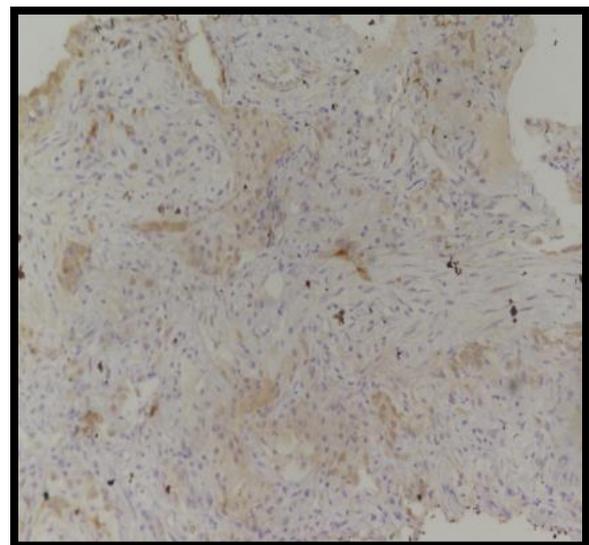


Fig 8: Shows S- 100 negative

Treatment

Treatment for Primary Pulmonary Hemangiopericytoma is Surgical resection. Depending on the Margin Status and Histopathology report. Adjuvant treatment should be added.

CONCLUSION

Primary Pulmonary Hemangiopericytoma in 64 year old female is very rare neoplasm. There is no uniform clinical or radiographic features. Mainly presents in older individuals as asymptomatic, non calcified mass on X ray. Tumor is mainly composed of spindle cells with prominent vacuolar channels. Its aggressive behavior cannot be predicted by histology. Tumor is very rare in incidence (<1%). A very few articles have been published on Pulmonary

Hemangiopericytoma worldwide (<50). Mostly this tumor is asymptomatic in its early stage and becomes symptomatic with advancing stage. Till date treatment approach of Primary Pulmonary Hemangiopericytoma is Surgical resection and depending on margins status different treatment can be considered. Adjuvant treatment is not clear as tumor is very rare in incidence. Prognosis of the tumor is still controversial.

So further reporting of such rare case is of para-amont importance to give the disease risk factor, prognosis and treatment.

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