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Pulmonary Epithelioid Hemangioendothelioma: A Case Report of Sleeve Resection of this Rare Lung Tumour

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Abstract **Case Report**

Reporting a 37 year old male with pulmonary epithelioid hemangioendothelioma (PEH), whom had undergone a sleeve resection of main left bronchus en bloc with the tumour. Presented with shortness of breath & was treated as pneumonia with left lung collapse. Bronchoscopy showed lesion at left main bronchus & unable to pass the obstruction. CT scan showed left main bronchus endobronchial lesion & histopathological reported it as suspected PEH. Mass had regrown 90% stenosis within 2 weeks. PET scan showed solitary disease at left endobronchial region. Left thoracotomy with sleeve resection of left bronchus en bloc with the tumour done. Intraoperative bronchoscopy showed endobronchial tumour located at the main left bronchus 1cm from carina. Histopathology confirmed the resected lesions are PEH. Patient recovered well. PEH is a histological diagnosis, recent reviews showed that radiological CT findings can be associated. No standard guideline treatment for PEH. There are cases reported with good response to combine chemotherapy and immunotherapy. Unilateral lesions, surgery is indicated. If lesion is positive for oestrogen and progesterone receptors, studies showed that hormonal therapy may benefit. Antivascular endothelial growth factor reported as part of treatment. Prognosis is unpredictable. In view of the rarity of the disease, multi-modulated approach is essential in the management and follow-up of the patient.

Keywords: pulmonary epithelioid hemangioendothelioma (PEH), endobronchial tumour, pneumonia, surgery.

INTRODUCTION

Pulmonary epithelioid hemangioendothelioma is a rare, low grade malignant vascular tumour [1]. Reporting a case of pulmonary epithelioid hemangioendothelioma, whom had undergone a sleeve Resection of Main Lt Bronchus en bloc with the tumour resection.

CASE

A 37 years old male, ex-smoker, with no family history of malignancy, initially presented with worsening shortness of breath, loss of weight and appetite, and was treated as pneumonia. He again presented few months later with similar complaint, and was again treated as pneumonia, but noted to have partial left lung collapse during this admission. He was subsequently referred to respiratory clinic. Initial bronchoscopy noted lesion at the left main bronchus and unable to pass through the obstruction. CT scan of the thorax showed left endobronchial lesion in left main bronchus (1.8x2.4x2.1cm) with left mediastinal shift and left lung lower lobe collapse.

Repeated rigid bronchoscopy and coring done. However, on surveillance bronchoscopy after 2 weeks, noted that mass had regrown with almost 90% stenosis of left main bronchus. Coring was also done. PET scan showed solitary disease at left endobronchial region with no metastasis. He was then referred to my centre for further surgical management. Left thoracotomy with sleeve resection of main left bronchus en bloc with the tumour done.

Intraoperative, bronchoscopy showed endobronchial tumour located at the main left bronchus around 1cm from carina covering >75% bronchus lumen, well capsulated with lobulated surface. No extraluminal extension. Histopathology examination showed the resected lesions consistent with epithelioid hemangioendothelioma, with clear margin distally and proximally. Patient was discharged home well, with a scheduled repeated CT scan.

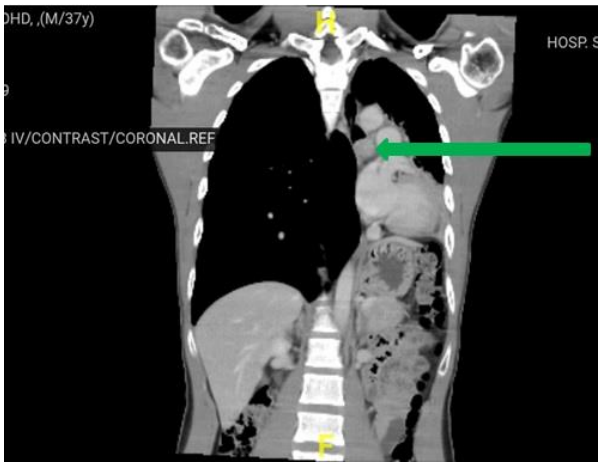

Figure 1: CT Thorax-coronal view, shows the mediastinal shift to the left side, and also the endobrochial lesion in left main bronchus-marked by green arrow

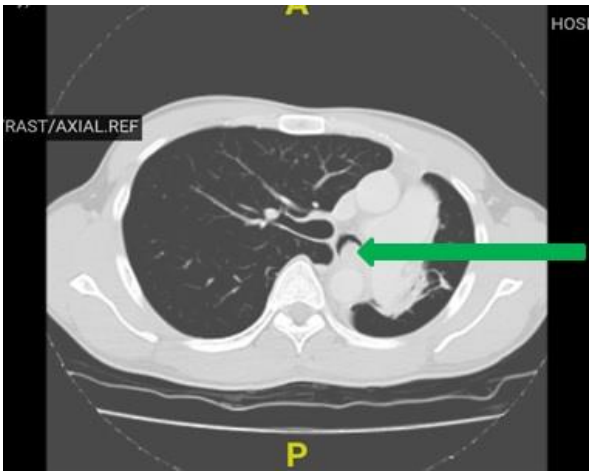

Figure 2: CT Thorax axial view: again shows the endobronchial lesion at the left main bronchus-marked in green arrow

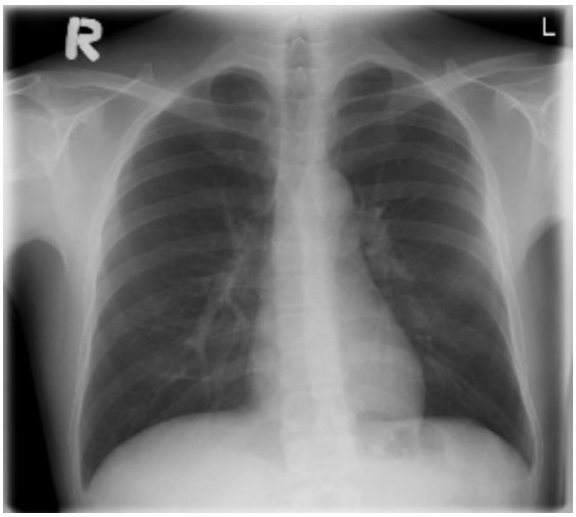

Figure 3: Chest Xray post operation shows left lung expanded with no more mediastinal shift

DISCUSSION

Discussing about the current literature review regarding the diagnostic, treatment and also prognosis of this disease. PEH is a histological diagnosis [2], but few recent reviews shows that, some radiological findings from CT scans can be associated with it [3,5], such as widespread nodules, with calcification, endobronchial mass with pneumonia changes, or a consolidation-like mass.

Generally, there are no single standard guidelines of treatment for PEH, but there are few cases that reported have responded to combine chemotherapy and immunotherapy [4]. In view of the surgical treatment, a review proposed surgery for unilateral lesion, but for bilateral multiple nodules, surgery is not indicated [1]. If the lesion is positive for oestrogen and progesterone receptors, it's documented that the patient may benefit from hormonal therapy. Few cases also documented to use anti vascular endothelial growth factor as part of the treatment, but the treatment option is not well established yet [4].

It is documented that the prognosis is very unpredictable. A review noted that the monoclonal antibody MIB-1 in the tumour cells, can be used as a prognostic factor, but further studies needed to establish this [4]. A close follow up, clinically and radiologically, could prove beneficial [5].

Compliance with Ethical Standards

I have of no conflicts of interest associated with this publication, and there has been no significant financial support for this work that could have influenced its outcome. No experimental procedures/research involving humans/animals had been conducted that is against the Human or Animal rights. The patient involved had given informed consent to publish his case.

Central Message

Rarity is a necessity. The rare nature of it, makes these diseases hard to manage and doesn't have a standardised universal approach of management, that is proven to be effective. Thus, I believe rare diseases such as Pulmonary Epithelioid Hemangioendothelioma is important to be discussed and I believe your journal would be a great platform for this.

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