CASE REPORT

Sclerosing Haemangioma of the Lung – An Unusual Multifocal Presentation

G Rosliza MBBCh; H Hairulfaizi MS; O S M Joanna MMed; D M Zamrin MS; A R M Ramzisham MS

Division of Cardiothoracic Surgery, Division of Cardiothoracic Anaesthesia, Heart and Lung Centre, Hospital Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, 56000 Cheras, Kuala Lumpur

SUMMARY

We present a rare case of a multifocal sclerosing haemangioma of the lung in a 49-year-old lady. A left pneumonectomy with complete excision of the tumour seems to be the curative treatment. The recent literature on this unusual presentation is reviewed.

KEY WORDS:
Sclerosing haemagioma, lung, pneumonectomy, thoracotomy

INTRODUCTION

Sclerosing Haemangioma (SH) of the lung is a rare benign tumour that occurs predominantly in middle-aged women. It was first described by Liebow and Hubbel in 1956[1]. Typically, it is a solitary and well-defined lesion. It comprises a mixture of four histologic patterns namely papillary, solid, sclerotic and haemorrhagic. Immunochemical analysis shows that SH is derived from pneumocytes type II[2]. We report a case of unusual presentation of pulmonary SH with multifocal appearance.

CASE REPORT

A 49-year-old lady, a chronic smoker, presented with an incidental finding of a mass at the left mid zone on chest radiograph. She declined further treatment until five years later when she presented with an episode of haemoptysis. Physical examination revealed decreased breath sounds in the left mid zone of the chest with dullness on percussion. A repeat chest radiograph showed a well-defined mass at the left mid-zone with only a slight increase in size as compared to her initial presentation. Computed tomography (CT) revealed a large mass at the left midzone measuring 8 x 10 x 9cm (Figure 1). It has well defined margins with heterogenous enhancement and specks of calcifications within the mass. Bronchoscopic examination and biopsy were inconclusive. Subsequently, she underwent a CT guided biopsy which was suggestive of vascular neoplasm.

She underwent a video assisted thoracoscopic assessment which revealed a large tumour occupying almost 80% of the left upper lobe with adherence to the lower lobe. There was parietal pleural infiltration of the tumour with three further nodules on the diaphragmatic surface, parietal pleura and anterior to the main lesion. Thoracotomy and left pneumonectomy was performed together with excision of the nodules.

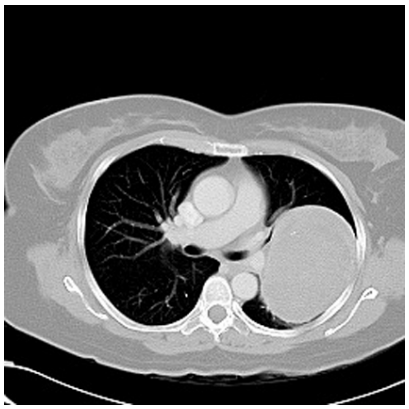

Fig. 1: CT showing a large intrathoracic mass.

Histopathological examination revealed a well-circumscribed spherical tumour with extensive necrotic and haemorrhagic areas (Figure 2). All the four histologic patterns of papillary, solid, sclerotic and haemorrhagic were present in the tissues derived both from the main lesion and the nodules. Immunohistochemically, the round stromal and surface cells showed a typical expression of thyroid transcription factor (TTF-1) and negative for CD34. Cytokeratin was positive for surface cells but negative for round stromal cells. Thus, the histological diagnosis of multifocal pulmonary sclerosing haemagioma was confirmed. Post-operative recovery was uneventful and she was discharged well on the 7th day following surgery. The patient has been followed up for the past two years and has been well in term of clinical and radiological examination.

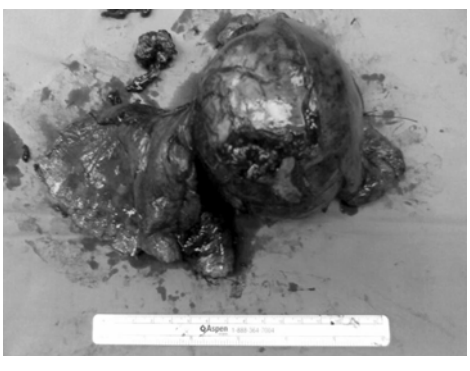

Fig. 2: Gross pathology of the specimen removed at surgery.

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Corresponding Author: Associate Professor Dr Mohd Ramzisham Abdul Rahman, Lecturer / Consultant Cardiothoracic Surgeon, Division of Cardiothoracic Surgery, Department of Surgery, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, 56000 Cheras, Kuala Lumpur Email: drramzi@ymail.com

DISCUSSION

Sclerosing haemangioma (SH) of the lung is a relatively rare tumour. It was first described by Liebow and Hubbell in 1956[1]. SH is believe to be a benign neoplasm. However, the histogenesis and clinicopathological features of this tumour remain controversial. SH presents most commonly in middle-aged females as asymptomatic, solitary, peripheral and well-circumscribed lesion. In this case, the lesion was discovered incidentally on plain chest radiograph and the patient remained asymptomatic for five years before her second presentation with haemoptysis. Symptomatic patients can also present with chronic cough or chest pain. The symptoms are due to enlargement of the tumour with bronchial infiltration or compression.

On gross examination, SH is solitary, well circumscribed and located in the peripheral lung with size ranging from 0.3-7cm in dimension. However, a rare case of multiple bilateral lesion has been reported[3]. In our patient, the atypical presentations of the lesions are multifocal and huge. The main lesion occupied 80% of the left upper lobe and adhered to the lower lobe. There were three other small nodules found on the diaphragmatic surface, pleura and anterior to the main tumour.

Characteristically, CT of pulmonary sclerosing haemangioma shows a mass with good enhancement. Sometimes, there is evidence of calcification within the mass, as observed in this patient. These characteristics are not specific for SH but are suggestive of a benign process. There are characteristic features of pulmonary SH such as, air trapping sign, tail sign, prominent pulmonary artery and intratumoural cystic area[4]. The CT features are correlated with histological findings. The differences in enhancement intensity correspond to the components of the tumour[4]. There are a few reports suggesting the role of positron emission tomography (PET) with 18[F]- fluorodeoxyglucose (FDG-PET) for the evaluation of SH but a large series is warranted before it can be used as a diagnostic tool.

Histologically, SH is characterised by two different cells types, surface cuboidal and stromal polygonal cells. Microscopic appearances of the lesion exhibit a mixture of four histologic patterns, namely papillary, solid, sclerotic and haemorrhagic. The proportions of the components vary and one of them tends to predominate. Immunohistochemically, both the surface cells and round stromal cells are positive for thyroid transcription factor-1 (TTF-1) and negative for CD34. Cytokeratin is positive for surface cells but negative for round stromal cells. It also has low mitotic index. On the basis of immunohistochemistry analysis, it originates from type II pneumocytes. All these features were demonstrated in this case. Despite its benign nature, the multifocal distribution of SH may suggest a low grade malignancy.

In general, SH is considered a benign lesion despite a few rare cases have been reported with regional lymph node metastases, which have no bearing on the prognosis. Hence, the definitive curative treatment for SH is a complete surgical excision. In conclusion, the atypical presentation of multifocal distribution of pulmonary SH does not affect the prognosis. The lesion has an excellent prognosis after complete surgical resection. Further effort should be made to elucidate the mechanism of multiplicity.

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