Letter to the Editor

Primary Epithelioid Angiosarcoma of the Lung Presenting as Left-sided Shoulder Pain

Dear Editor,

Primary angiosarcoma of the lung is a rare, often difficult to diagnose disorder with non-specific respiratory manifestations. Angiosarcoma involving the lung is usually a result of metastatic disease. Among the commonest presentation of this rare disease is pulmonary haemorrhage, which has been described in a handful of cases.[1,2] We report a unique presentation of primary pulmonary angiosarcoma in a 23-year-old man who presented with a metastatic left-sided shoulder and chest pain. The diagnosis was confirmed by a computer tomography (CT)-guided biopsy of the left lung mass. Immunohistochemical studies were positive for Vimentin, CD34, CD31 and Factor VIII and negative for CK, CK7, epithelial membrane antigen (EMA), thyroid transcription factor 1 (TTF1), Calretinin, smooth muscle actin (SMA), Desmin, leukocyte common antigen (LCA) and human melanoma black (HMB)45.

Case Report

A 23-year-old man, a non-smoker with no significant prior medical illness presented with a 6-month history of left-sided shoulder and left chest pain. He was initially treated for muscular pain in another hospital but as the pain worsened, he was admitted to our centre for further investigation. He experienced shortness of breath on exertion and weight loss. There was no history of prolonged fever, cough nor exposure to tuberculosis. He had finger clubbing and a left sided pleural effusion. There were no signs of lymphadenopathy and hepatosplenomegaly. The laboratory investigation results were normal. Chest radiograph showed a left pleural effusion with consolidation at the lingula area. Pleural fluid was haemorrhagic and exudative by Light's criteria. The cytology results were negative for malignant cells.

Investigations for pulmonary tuberculosis and connective tissue disease were all negative. The initially diagnosis was pneumonia with parapenumonic effusion. However, he failed to respond to antibiotic therapy and was subsequently started on empirical antituberculous therapy. A pleuroscopy was attempted but failed due to the narrowed rib spaces; however a 12F chest tube was inserted which removed the haemorrhagic fluid.

A CT scan of the thorax revealed a large left pleural effusion with pleural thickening and collapse of the left lung parenchyma. A heterogenous enhancing mass measuring 5.0 x 3.3 x 4.1 cm was found within the collapsed lung (Fig. 1). The mass encased the left descending pulmonary artery and there was ipsilateral hilar lymphadenopathy. The left hemithorax was smaller indicating a chronic process possibly tuberculosis or a mitotic process. This was thought to be unrelated to the current diagnosis. There was also evidence of multiple skeletal metastases in particular at the level of C6 with destruction of the vertebrae. A CT guided biopsy of the left lung consolidation was done. The histopathological examination (HPE) results revealed an epitheliod angiosarcoma of the lung. Immunohistochemical studies were positive for CD34 (Fig. 2), Vimentin, CD31 and Factor VIII and negative for CK, CK7, EMA, TTF1, Calretinin, SMA, Desmin, LCA and HMB45. A bone scan confirmed multifocal metastases. Despite initiation of chemotherapy, the patient died 2 weeks after the treatment.

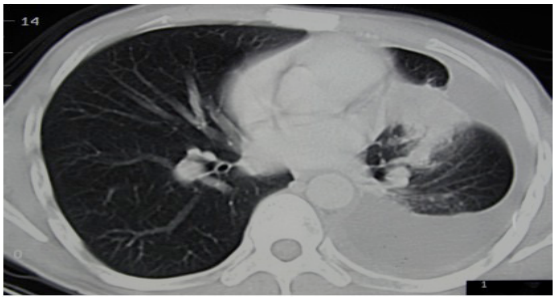

Fig. 1. CT scan of the chest showing left sided pleural effusion with enhancing pleural and heterogenous mass in the left lung.

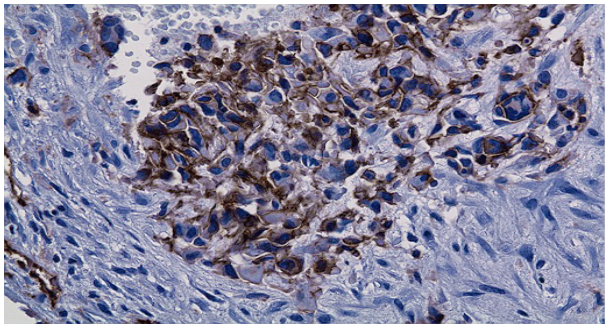

Fig. 2. Lung biopsy (X400) Cells stain positive for CD 31.

CONCLUSION

Angiosarcomas are rare and represent less than 2 percent of all sarcomas. The most frequently involved primary sites are heart, liver and breast with over 100 cases of each reported.[3] Angiosarcomas of the lungs are usually metastasis. Primary angiosarcoma of the lung is extremely rare and is usually diagnosed quite late due to low index of suspicion. In Malaysia, pulmonary tuberculosis is a common cause of haemorrhagic pleural effusion. As a result of this, our patient was treated empirically for pulmonary tuberculosis (PTB).

Patients with primary angiosarcoma can present with chest pain, haemoptysis (which can be massive), dyspnoea, cough and weight loss. Our patient presented with left shoulder pain, which is most likely, a referred pain from compression of the nerve root at the cervical spine. His CT thorax showed destruction of cervical vertebrae in particular C6. The bone scan did not show any metastasis to the left shoulder joint. Reported chest radiograph findings have ranged from normal to multiple nodular densities with or without pleural effusions to diffuse alveolar infiltrates compatible with pulmonary haemorrhage. Metastatic angiosarcoma lung lesions are commonly multiple. There have been reported as solid nodular or thin-walled cysts, with haemorrhagic change.[4] This was not seen in our patient. Based on the CT thorax findings, we assumed that this patient had a primary pulmonary angiosarcoma.

The HPE was consistent with an epithelioid angiosarcoma and this was also supported by positive reaction for factor VIII-related antigen, CD34 and CD31, which are specific markers for tumours derived from the endothelium. Predisposing factors for angiosarcomas include polyvinyl chloride and thorium dioxide exposure, postmastectomy and postirradiation states (cutaneous angiosarcoma) and chronic empyema for pleural space angiosarcomas.[5] Our patient had none of these.

Prognosis of pulmonary angiosarcoma has been shown to be poor with almost all patients dead within months of initial presentation. Our patient died within 4 months from presentation of the left shoulder pain. We would like this case report to be added on to the current list of atypical presentation of primary pulmonary angiosarcoma. We would also like to highlight the misdiagnosis of pulmonary tuberculosis in this patient. The presenting complaint of shoulder pain was disregarded in favour of the more obvious haemorrhagic pleural effusion. We believe the hunt for rarer causes of haemorrhagic pleural effusion might have led to an earlier diagnosis.

Acknowledgements

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Wan Rosmaiza Wan Musa,[1] *MBChB (Leicester),* Mohamed Abdulwakil Elraied,[2] *MB Bch (Tripoli, Libya),* Koon Seng Phang,[2] *MBBS (MU) DCP (London),* Yew Gee Kwah,[3] *MBCHB (Dundee,UK),* Sook Pei Tan,[3] *MD(UKM), MMED(UKM),* Roslan Harun,[1] *MD (UKM), MRCP (UK), PHD (Molecular Biology Leeds),* Andrea YL Ban,[1] *Mb Bch Bao (Ireland), M.MED (UKM)*

[1]Dept of Medicine, Pusat Perubatan University Kebangsaan Malaysia
[2] Dept of Pathology, Pusat Perubatan University Kebangsaan Malaysia
[3]Dept of Radiology, Pusat Perubatan University Kebangsaan Malaysia

Address for Correspondence: Dr Andrea Yu-Lin Ban, Department of Medicine, Pusat Perubatan Universiti Kebangsaan Malaysia, Jalan Yaakob Latif, Bandar Tun Razak, Cheras, 56000, Kuala Lumpur, Malaysia.
Email: andreaban@gmail.com