

Case report:

Pleuropulmonary Solitary Fibrous Tumour with Paraneoplastic Syndrome

Ummu Afeera Zainulabid¹, Megat Razeem Abdul Razak², Nor Hafiza Md Salleh³, Noriah Othman⁴, Dahlia Mohamed⁵

Abstract:

The pleuropulmonary solitary fibrous tumour (SFT) is a rare type of tumour. This paper outlined a 63-year-old female who came to the hospital with two weeks history of chronic cough, shortness of breath, and hypoglycemia. Contrast-Enhanced CT Thorax showed a huge heterogeneously-enhancing mass occupying the right hemithorax. US-guided biopsy followed by histological examination showed the features of an SFT. In view of the association between pleuropulmonary SFT and hypoglycaemia, the patient was highly likely to be suffering from a paraneoplastic syndrome known as Doege-Potter Syndrome.

Keywords: Solitary fibrous tumour, Doege Potter Syndrome, superior vena cava obstruction.

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Introduction

The pleuropulmonary solitary fibrous tumour (SFT) often follows a silent clinical course until the tumour size is large enough to produce compressive symptoms. In certain cases, SFT presents with paraneoplastic syndromes one of the most commonly encountered being the Doege-Potter syndrome. Doege-Potter syndrome is a paraneoplastic syndrome that presented with hypoglycaemia as a result of non-islet cell tumours like SFT.

Case Report

A 63-year-old female, non-smoker, non-diabetic initially presented to a district hospital with a two-week history of cough associated with difficulty in breathing and constitutional symptoms. Clinical examination revealed an elderly cachexic lady with a tracheal shift to the left side, reduced breath sound and dullness to percussion in the right lung. Other examinations were unremarkable.

The chest radiograph showed a homogenous opacity obscuring the right hemithorax. There was also a mediastinal shift with tracheal deviation to the left. Contrast-Enhanced CT Thorax showed a

huge heterogeneously-enhancing mass occupying the right hemithorax. It measured approximately 16.4 cm x 16.5 cm x 21.1 cm. Minimal residual collapsed lung was noted in the right apical region. Pleural tapping was done at the district hospital. The pleural fluid was exudative in nature. The patient was also found to have unexplained hypoglycemia. She was initially thought to have adrenal metastasis from the lung and started on IV Hydrocortisone 100mg TDS. However, the hypoglycemia persisted. Her condition worsened and she was transferred to our care in the tertiary hospital. She was then put on non-invasive ventilation. She was found to have superior vena cava syndrome (SVCS) as evidenced by the venous distention in the neck, upper chest, and arms as well as facial swelling. In view of that, she was immediately started on high dose IV Dexamethasone. Following that, the SVCS and refractory hypoglycemia improved remarkably.

Once her condition improved, a trucut biopsy of right lung mass was done under ultrasound guidance. Microscopically, the section showed spindle to oval-shaped tumour cells. The

1. Internal Medicine Specialist & Medical Lecturer, Department of Internal Medicine, Kulliyah of Medicine, International Islamic University of Malaysia, Kuantan, Malaysia.
2. Pulmonologist, Tengku Ampuan Afzan Hospital, Kuantan, Malaysia.
3. Pathologist, Tengku Ampuan Afzan Hospital, Kuantan, Malaysia.
4. Pathologist, Serdang Hospital, Kajang, Selangor, Malaysia.
5. Radiologist, Sultan Haji Ahmad Shah Hospital, Temerloh, Pahang, Malaysia.

Correspondence to: Dr. Ummu Afeera Zainulabid, Internal Medicine Specialist & Medical Lecturer, Department of Internal Medicine, Kulliyah of Medicine, International Islamic University of Malaysia, Kuantan, Malaysia. E-mail: ummuafeera@iiium.edu.my

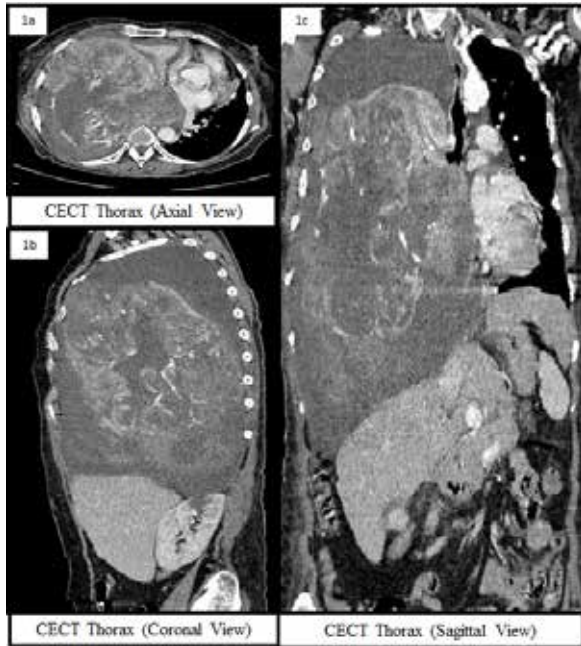


Figure 1: Contrast-enhanced CT Thorax in axial, coronal, and sagittal images show a huge heterogeneously-enhancing mass occupying the right hemithorax with massive right pleural effusion.

hypocellular and hypercellular areas were seen within a ropey collagenous stroma. Pericytic vascular patterns were also visible. The tumour cells displayed round to oval nuclei, mild to moderate nuclear atypia, and pale indistinct cytoplasm. Occasional multinucleated or large atypical tumour cells were present. Mitotic figures were present and seen (seven mitoses per 2mm). No marked nuclear pleomorphism or bizarre tumour cells were noted. There was no necrosis or haemorrhage.

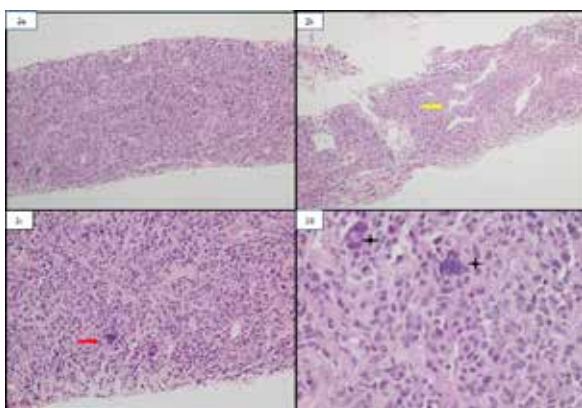


Figure 2: a) The biopsy shows spindle to oval-shaped tumour-H&E 100x magnification. b) Pericytic vascular pattern (yellow arrow)-H&E 100x magnification. c) Multinucleated tumour cells (red arrow)-H&E 200x magnification. d) Multinucleated tumour cells (star)-H&E 400x magnification.

The tumour cells were positive for CD34, BCL2, CD99, and STAT 6 with patchy CK AE1/AE3 positivity. Based on the histological findings, a diagnosis of a solitary fibrous tumour (SFT) was reached. They were negative for CK7, EMA, and TTF1. The patient was referred to the national oncology centre in Hospital Kuala Lumpur for the continuation of care.

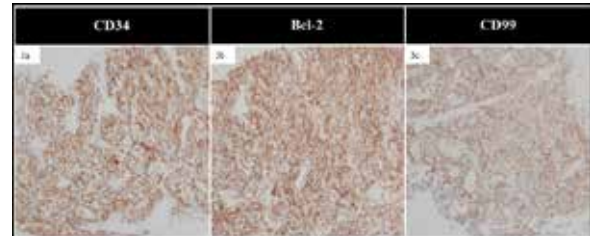


Figure 3: Immunohistochemistry for CD34, Bcl-2, and CD99 are positive – 200x magnification.

Discussion

SFT was first described in 1931. It has been described with various nomenclatures, including solitary fibrous mesothelioma, pleural fibroma, and others. SFT may present in patients of all age groups but it is most commonly encountered among those aged 50-70 years.¹

In this case report, the patient was found to have a very large SFT in the right lung. The lesion occupied the right hemithorax, leading to massive right pleural effusion. Intrathoracic SFT may arise in the pleura, mediastinum, or lung parenchyma. Aside from the pleura, SFT has also been found in the serosal membranes, the dura of the meninges, and deep soft tissues.²

It was reported in the literature that pleuropulmonary SFT often presents with non-specific pulmonary symptoms such as cough, shortness of breath, or chest pain. The patient in this case report presented with similar symptoms.¹ Macroscopically, pleuropulmonary SFT usually appears as a well-delineated and occasionally lobulated mass of soft tissue attenuation arising from the pleura.³ Histologically, SFT is usually described as a low-grade neoplasm with variable cellularity. The cancer cells may present in oval or fusiform shape with oval nuclei and well-distributed chromatin. Recently, immunohistochemistry has gained importance as an extremely useful tool for the diagnosis of SFT. Perrot et al. summarised the most essential immunohistochemical characteristics in SFT as positive for vimentin and negative for keratin. Furthermore, CD34 is positive in the majority of the benign SFTs as well as some malignant ones.

However, CD34 remains negative for most of the other tumours of the lung.⁴

There are a few criteria that suggested malignancy based on this patient's biopsy, namely increased mitotic activity (> 4/2 mm), hypercellularity, and moderate nuclear atypia. However, other features for malignancy i.e. tumour necrosis or haemorrhage were not seen. The infiltrative nature of the tumour could not be ascertained histologically. Differentiating between benign and malignant tumors is challenging. However, there are a few radiological characteristics that are highly suggestive of malignancy, including lesions that are larger than 10 cm with central necrosis and large pleural effusions. All these features were seen in this case.⁴

Furthermore, this patient had a unique presentation of non-insulin mediated hypoglycemia. SFT is a known tumour that causes non-insulin mediated hypoglycaemia. This is a type of paraneoplastic syndrome known as Doege-Potter syndrome. It occurs in less than 5 percent of SFT. It is primarily seen in large peritoneal/pleural tumours such as this case. It happens as a result of tumour secretion of large insulin-like growth factor II (IGF2). Fortunately, it responded well to high dose

glucocorticoid.^{4,5}

Conclusion

Pleuropulmonary SFT is a rare condition. Highly-specific physical, imaging, and histopathological examination are needed to diagnose the condition. However, the treatment of pleuropulmonary SFT is not well-established at this point in time, thus long-term follow up is mandatory. This case report emphasised the importance of recognising Doege-Potter syndrome in a patient with SFT and hypoglycemia.

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