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Primary Pulmonary Synovial Sarcoma; A Case Report of a Large Lung Mass

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Abstract | **Case Report**

Synovial sarcoma is a rare soft tissue sarcoma which arises from immature mesenchymal tissue and has been described to occur in close relation to tendons, tendon sheaths and bursal structures. Synovial sarcoma can also occur in the lung, which is known to be part of a very rare subset of primary lung malignancies. There is no standard management for primary pulmonary synovial sarcoma (PPSS) but it is recommended that resection is done as early as possible. We describe a case of a 37-year-old male with a large primary pulmonary synovial sarcoma (PPSS). This report includes the diagnostic process and the adopted treatment approach for this patient.

Keywords: Primary Pulmonary Synovial Sarcoma (PPSS), Synovial Sarcoma, Lung Cancer, Rare Tumor, Surgical Resection.

INTRODUCTION

Synovial sarcoma is a rare soft tissue sarcoma which accounts for 8% of all soft tissue tumours [1-3]. It has been described to occur in locations unrelated to synovial tissue which include the lung. Primary pulmonary synovial sarcoma (PPSS) is a rare pulmonary disease which comprises of only 0.5% of all lung malignancies [2-4.] This disease usually has a slow and insidious growth and tends to be large in size upon detection and diagnosis. We report a 37-year-old male with a large 12 cm PPSS which was successfully resected.

CASE REPORT

A 37-year-old male, who is a non-smoker with no significant past medical history presented with two months history of persistent cough despite empirical treatment with antibiotics. Subsequently, a chest X-ray was done which showed a large right opacity (Fig. 1). He subsequently underwent a CT Thorax which showed a large heterogenous mass of size 11cm x 10cm x 12.1cm (Fig. 2). He proceeded to undergo bronchoscopy which found RB6 was compressed (Fig.3). Bronchial biopsy was performed and the histopathology showed 'Monophasic Synovial Sarcoma (TLE +ve, Vimentin +ve, CKAE1/AE3 -ve, SMA -ve, Desmin -ve, CD34 -ve, and S100 -ve).'

Patient further underwent PET CT scan (Fig 4) which showed large intense FDG-avid lesion in the right lower lobe. This appear to be involving the right oblique fissure an adjacent right pleura; cT4N2M0.

A staging MRI brain was done which didn't detect any cerebral metastases. His spirometry assessment was normal and his preoperative bloods were unremarkable.

Patient underwent intraoperative bronchoscopy which showed no intraluminal lesion, but indentation could be seen on the membranous portion of bronchus intermedius. He subsequently underwent a right thoracotomy, right lower lobectomy with extra-pleural and lymph node dissections.

Primary intraoperative finding revealed a 12cm right lower lobe tumour which was adherent to the chest wall posteriorly causing anatomical distortion to the right lower lobe pulmonary artery and the middle lobe pulmonary artery. Post-operatively, he progressed well with his chest tube uneventfully removed at POD 3. He was discharged home at POD 4.

Final histology of the resected right lower lobe, pleural nodule and lymph node tissues showed:

1. Right lung lower lobe: Monophasic synovial sarcoma, FNCLCC grade 3 (score 7/8),

confined within lower lobe, pT4, completely excised.
2. Pleural nodule: Granulation tissue nodule with congestion and oedema, no malignancy seen.
3. Station 2, 4,7, 9 and 11 showed no metastasis
4. Final staging: pT4, pN0, M0.

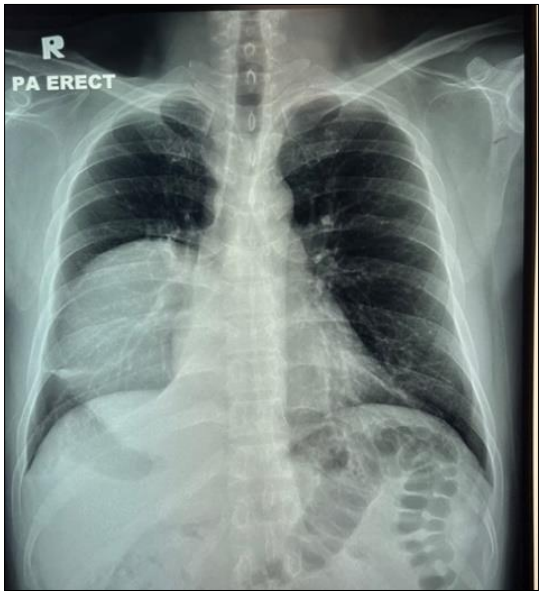

Fig. 1

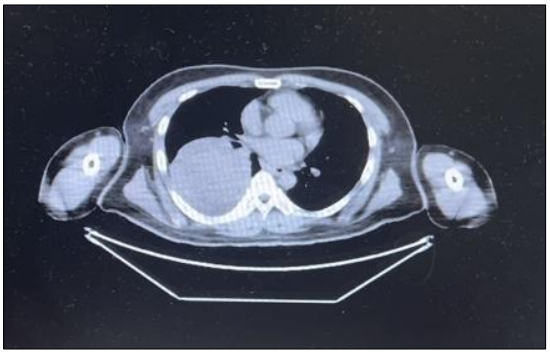

Fig. 2

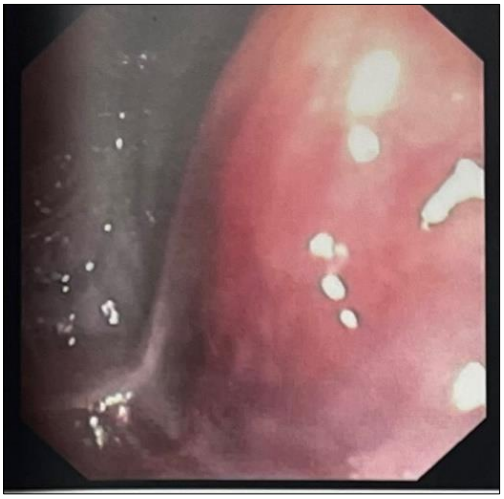

Fig. 3

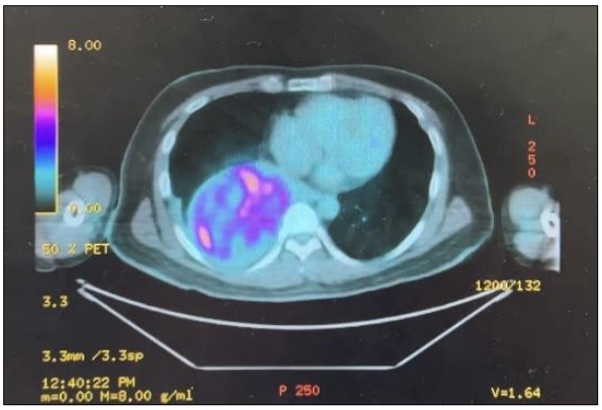

Fig. 4

DISCUSSION

Synovial sarcoma is considered to be a rare soft tissue sarcoma which accounts for 8% of all soft tissue tumours in the body [1-3]. It is mistakenly thought to arise from the synovium as it most commonly occurs in the extremities, especially approximate to large joints [1]. It has been described to occur in close relation to tendons, tendon sheaths and bursal structures [4]. However, it has also been described in locations unrelated to synovial tissues. The synovial subset of primary pulmonary synovial sarcoma (PPSS) is a rare disease which comprises only 0.5% of all primary lung malignancies [2-4]. Sarcomas in the pulmonary system are most commonly secondary to extrapulmonary sarcomas [3].

PPSS affects both genders equally [5, 6]. It occurs mainly in adolescents and young adults [1]. Most patients present between the age of 15 and 35 years old, with 90% of the cases are reported before the age of 50 years [5]. It has been reported that the average age at presentation is 25 years old [2]. The incidence of either right or left PPSS is similar [5], [3], however it is most commonly seen at the apex of the affected lung [3].

Synovial sarcoma is a rather a misnomer as it is not derived from synovium (1),(2). It is derived from immature mesenchymal tissues [2], such as skeletal and smooth muscles, adipose tissues, bone, cartilage, and synovial tissues [3]. There is no synovial tissue located in the pulmonary system. The tumour arises from pluripotent mesenchymal cells which then differentiate into synovial tissue [3]. Most PPPS are located in the lung parenchyma and it is uncommon for it to extend into the bronchial tree or occur in the heart or pericardium [7].

PPSS grows slowly and insidiously, and most of the time this often led to delay in diagnosis and definitive therapy [3]. Clinical features and presentation of the patients are mainly dependent on the size as well as location of the tumour. The common symptoms are chest pain, cough, dyspnea and hemoptysis [5]. In some cases, patients may present with pleural or pericardial effusions, recurrent pneumothorax or endobronchial

mass [5]. PPSS tends to be large and may cause partial or complete opacification of the affected lung [3]. Most tumours larger than 7cm are reported to cause compression of the surrounding tissue [8].

A chest X-ray of PPSS will typically show a well-defined uniform homogenous mass lesion with round, lobulated borders. CT scan of PPSS is similar to those of many common pulmonary and pleural neoplasms of the lung and pleura, including primary and metastatic lung cancer [9]. It will typically show heterogenous enhancement due to presence of a necrotic, cystic or friable portion of the tumour interspersed with a solid enhancing component of the tumour [3].

Immunostaining also plays an important role in diagnosis. Monophasic synovial sarcomas usually stain positive for vimentin, Bcl-2, CD-99, TLE, FLi-1, and S-100 [3]. Our patient exhibits a positive TLE and vimentin.

There is no standard, universal recommendation for the optimal management of patients with primary pulmonary synovial sarcoma [3]. However, it has been reported that the current treatment for PPSS is surgical resection followed by chemotherapy, radiation therapy or combination of these two modalities [6]. Thus, resection of the tumour should be done as early as possible. Synovial sarcoma is relatively sensitive to chemotherapy particularly to adriamycin alone or in combination with ifosfamide [10].

PPSS has a relatively poor prognosis with an overall 5-year survival rate of 50% [2-4]. There are several factors that predict worser prognosis which include tumour size (>5 cm), male gender, older age (>20 years), extensive tumour necrosis, high grade lesion, large number of mitotic figures (>10 per 10 high-powered fields), evidence of neurovascular invasion, and, recently, the *SYT-SSX1* variant [11]. However, the fundamental prognostic factor is the ability to achieve complete resection [2-6].

CONCLUSION

PPSS is a rare pulmonary tumour which is mainly seen in young adults. It has a slow and insidious growth which often results in delayed diagnosis and treatment. PPSS has similar findings to those of many common pulmonary and pleural neoplasms of the lung and pleura on CT scan. Histopathological confirmation including immunostaining information from biopsied or resected tissues are important tools in clinching the diagnosis. There is no standard management approach of patient with PPSS, however, it is recommended that resection of tumour is done as early as possible followed by chemo-radiotherapy. The ability to achieve complete resection remains the main favourable prognostic factor for this condition.

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