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Case Report

Primary pleuropulmonary synovial sarcoma mimicking a carcinoid tumor: Case report and literature review

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ABSTRACT

Primary pleuropulmonary synovial sarcoma is a rare malignancy. Commonly described radiologic features in the literature include pleural disease and/or effusion, lack of calcification and high uptake on positron emission tomography computerised tomography. A 68-year-old woman presented with a 3-month history of cough. Imaging studies showed a right upper lobe mass with internal foci of calcification, endobronchial extension, and low fluorodeoxyglucose avidity on positron emission tomography computerised tomography, leading to an initial diagnosis of carcinoid tumor. However, histologic specimens suggested an unexpected diagnosis of aggressive synovial sarcoma, and the case was referred to the sarcoma MDT. Metastatic synovial sarcoma was ruled out, and radical surgical excision of the lesion was performed. This article highlights the multiple atypical features of primary pleuropulmonary synovial sarcoma as seen in this case and reviews imaging findings described in the literature. Radiologists should be aware of this unusual yet aggressive type of sarcoma.

Introduction

Primary lung synovial sarcoma is an aggressive malignancy that is rarely encountered in routine practice and within the Sarcoma multidisciplinary team (MDT), with only a small number of reported cases in the literature. Nevertheless, radiologists need to be aware of the imaging findings, differential diagnoses and management pathway of this entity, as early diagnosis and complete surgical resection are the most important prognostic factors. Radiology plays an important role in suggesting the diagnosis, ruling out the most important differential diagnosis of metastatic synovial sarcoma and following up the patient. We present a rare case of primary pleuropulmonary synovial sarcoma (PPSS) that was misdiagnosed initially as a carcinoid tumor. We will summarize the literature findings about primary lung synovial sarcoma emphasizing the pertinent radiologic features and compare them to the atypical features seen in our case. The aim of this article is to familiarize the reader with the radiologic features and imaging pitfalls of this rare malignancy, as early

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recognition and complete surgical resection are crucial for the best outcome.

Case report

A 68-year-old woman presented to the Chest clinic with a persistent cough of 3-month duration. There was no history of hemoptysis, smoking, or other significant medical history. Clinical examination was unremarkable. A chest radiograph (Fig. 1) showed a well-defined mass in the right upper zone. Subsequently, she had an urgent contrast-enhanced CT scan of the chest, abdomen, and pelvis (Figs. 2 and 3) confirming a 5.6 × 5.4 × 5-cm soft tissue mass in the right upper lobe with an endobronchial component filling the right upper lobe bronchus extending approximately 1 cm distal to the carina. The lesion demonstrated mainly soft-tissue attenuation (42-50 Hounsfield units) with internal specks of course calcification. There was secondary atelectasis of the anterior segment of the right upper lobe. No enlarged thoracic lymph nodes or other lesions were seen. The visualized skeleton was normal. Positron emission tomography computerised tomography (Fig. 4) showed low fluorodeoxyglucose (FDG) activity with a maximum standardized uptake value of 3.8 within the lesion. No other areas of abnormal uptake were present. The initial clinical and radiologic impressions were pulmonary carcinoid tumor. However, histologic assessment of a transbronchial biopsy suggested a synovial sarcoma. The case was discussed at both the lung and soft-tissue sarcoma MDT, and the concluding diagnosis was PPSS. The patient underwent complete surgical excision. The surgical specimen showed grade 3 PPSS with negative resection margins. The patient had an uneventful recovery and is currently being followed up with 6-monthly CT scans. Adjuvant therapy was considered unnecessary in view of the complete surgical resection.

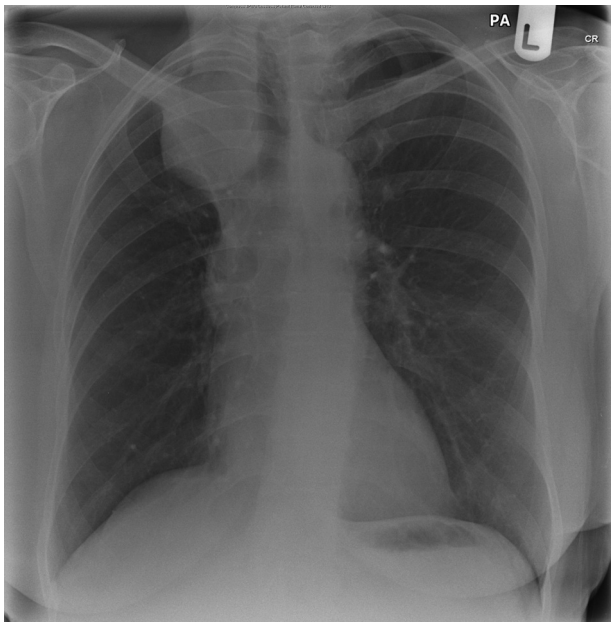

Fig. 1 – PPSS in a 68-year-old woman. Posteroanterior chest radiograph demonstrating right upper zone well defined mass abutting the right trachea. There is volume loss of the right upper lobe with tracheal shift to the right.

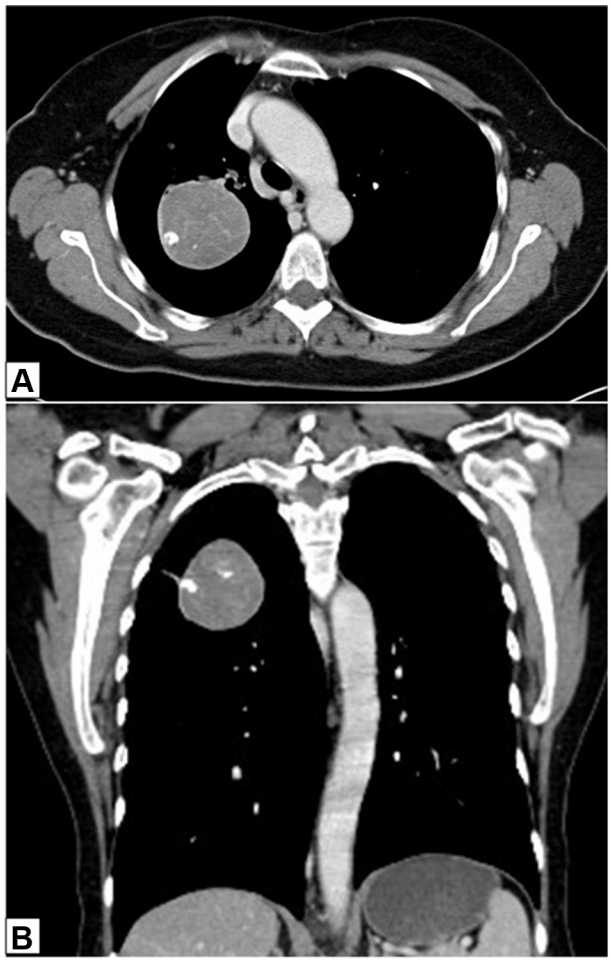

Fig. 2 – PPSS in a 68-year-old woman. (A) Axial contrast-enhanced CT scan of the thorax demonstrating a well defined, heterogeneous mass in the right upper lobe with internal septation, and peripheral thin enhancing rim. (B) Coronal-reconstructed image shows multiple well-defined foci of calcification.

Discussion

Synovial sarcoma is a type of spindle cell tumor accounting for 2.5%-10% of all soft-tissue sarcomas. It is primarily seen in a para-articular location within the extremities in adolescents and young adults, with a slight predilection for the knee joint. Rare sites include the head and neck, mediastinum, lung, pleura, and chest wall [1,2]. Primary pulmonary sarcoma is rare constituting approximately 0.5% of primary lung malignancies [2,3]. The synovial subset of primary lung sarcoma is very rare with a limited number of reported cases in literature. It is often difficult to determine the exact site of the primary tumor in terms of whether it is pleural or parenchymal, and therefore, it is often referred to as PPSS. PPSS usually affects people in the 4th or 5th decade of life [1,4,5] with a reported