Case Report

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

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A R T I C L E  I N F O

Article history:
Received 24 January 2016
Accepted 6 February 2016
Available online 19 March 2016

Keywords:
Primary pleuropulmonary synovial sarcoma
Synovial sarcoma

A B S T R A C T

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.

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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...
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 coarse 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.

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

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.
age ranged between 9 and 77 years [5]. The literature either
describes no specific sex predilection [2,5,6] or a slight male
predominance [1,3,7]. Clinical symptoms include dyspnea,
cough, chest pain, and hemoptysis [1,2,5,6,8]. No definite risk
factors have been established, although there is a case report
of PPSS after radioactive iodine therapy for thyroid cancer [9].
No relation to cigarette smoking has been established [7].

PPSS is a mesenchymal tissue cell tumor. Macroscopically,
it consists of uniform or nodular solid soft-tissue component
intermingled with variable necrotic and hemorrhagic compo-
nents. Surrounding pleural adhesions and a capsule are
described in few cases [2]. Microscopically, it has 4 histologic
subtypes including monophasic fibrous (spindle), monophasic
epithelial, biphasic, and poorly differentiated. Monophasic

subtypes are the most common [1,2,7,10]. Immunohisto-
chemistry is positive for Bcl-2 protein in approximately 79% of
cases [2,10], and cytogenetic analysis demonstrates a char-
acteristic chromosomal translocation t(x;18)(p11;q110) in 90%
of cases [1,2,8]. Historically, the nomenclature of synovial
sarcomas is because of the impression of associated syno-
vium, with the epithelial components representing synovial
slits [4]. However, these neoplasms have no established rela-
tionship to synovial tissue, making the terminology of “sy-
novial sarcoma” a misnomer [2].

Although PPSS is often detected on a chest radiograph, the
plain radiographic findings are not specific and indistinguish-
able from other primary lung tumors [5]. Plain radiographic
detection and appearance depend on the site and size of the
lesion. Although distinction between primary parenchymal or
pleural disease can be difficult, parenchymal predominant
synovial sarcoma will usually manifest as a well defined,
rounded mass, or less commonly as a consolidation with rather
ill-defined margins. Pleural synovial sarcoma is usually seen as
a pleural-based mass or an area of pleural thickening [1,2,5]. An
associated ipsilateral pleural effusion is very common and
described in multiple cases [1,2,5] and may be related to acute
or recurrent hemotherax [1,11]. Pneumothorax is uncommon
and only seen in a few cases [5,11]. Interestingly, our case did
not demonstrate any pleural effusion. Large tumors can cause
significant or complete opacification of the involved lung and
mediastinal shift [2,5]. In a review of 5 cases, the smallest
tumor measurements were 6 × 5 × 6 cm, and the largest tumor
measurements were 14 × 15 × 19 cm [1]. The range of the
maximum measurement of tumors in a review of 12 cases was
5-20 cm with an average value of 10 cm [2].

Fig. 3 — PPSS in a 68-year-old woman. (A) Axial contrast-
enhanced CT scan of the thorax (lung window) showing
right upper lobe bronchus endobronchial tumor extension
and associated right upper lobe anterior segment
atelectasis. (B) Sagittal-reconstructed image (lung window)
showing the position of the tumor in the posterior segment
of right upper lobe abutting the oblique fissure with
atelectasis of the anterior segment of right upper lobe.

Fig. 4 — PPSS in a 68-year-old woman. (A, B) Fused axial
images of positron emission tomography computerised
tomography scan showing low fluorodeoxyglucose avidity of
the right upper lobe tumor with endobronchial extension and
a focus of calcification.
PPSS is usually seen as a heterogeneous mass on non-enhanced CT because of a combination of solid and cystic components. Contrast-enhanced CT will show a heterogeneous enhancement pattern usually involving the nodular soft tissue, and a nonenhancing, cystic and/or necrotic component. A thin, irregular enhancing peripheral rim is also a common feature [1,2,4,11]. Septation is uncommon and only described in 2 cases [2]. A surrounding rim of ground glass changes can be seen on CT [5,12]. Our case appeared as a heterogeneous lesion with multiple thin enhancing septae.

Internal calcification apparent on imaging is very rare and described only in one case [13]. A separate case reported calcification that was only visible on the gross specimen after surgical excision but not radiologically [2]. Multiple foci of calcification, which led to the initial diagnosis of carcinoid tumor, are seen in the presented case. Although internal calcification is rare, it can be seen in PPSS, and hence, the diagnosis should not be excluded. It is worth mentioning that calcification is often present in metastatic synovial lung carcinoma, which is the most important differential diagnosis considering the lungs are involved in 94% of metastatic synovial sarcomas [14].

The tumor does not have a specific predilection for a certain lung lobe. However, upper lobe lesions were common in one case series [3]. Metastasis is usually hematogenous rather than lymphatic and enlarged thoracic lymph nodes secondary to disease involvement are extremely rare. To our knowledge, there is only one reported case of PPSS with lymph node metastasis, which was in a paraesophageal location [15].

Metastatic pleural nodular disease and bone involvement can be seen [2,14]. However, obvious chest wall muscular involvement or bony cortical destruction favors a synovial sarcoma arising from the chest wall [5,14].

Although positron emission tomography computerised tomography imaging often shows increase uptake within the lesion [2,5], our case only demonstrated low fluorodeoxyglucose avidity mimicking a carcinoid tumor.

Magnetic resonance imaging usually shows a heterogeneous tumor that is predominately low to intermediate signal on T1-weighted images and high signal on T2-weighted images [2,4]. Fluid–fluid levels due to hemorrhage or necrosis are also reported [6]. Gadolinium administration usually shows heterogeneous internal and peripheral rim enhancement [2,4].

The most important differential diagnosis is metastatic synovial sarcoma as the lung is the most common site for metastasis, affected in 94% of metastatic cases [2,14]. Since both primary and metastatic lung synovial sarcoma will have similar histologic and cytogenetic findings, radiology and clinical assessment play the most important role in differentiation between the two [2]. Other differentials include primary and metastatic lung neoplasms, localized pleural fibrous tumor, mesothelioma in cases of pleural disease, and other rare primary parenchymal sarcomas. The presence of hilar or mediastinal enlarged lymph nodes suggests a diagnosis other than PPSS [2,5].

PPSS is more aggressive than soft-tissue synovial sarcoma, possibly because of a relative larger size and later presentation [16] with reported survival figures of 6-58 months from diagnosis [3,5]. Factors indicating good outcome are tumor size <5 cm, low mitotic rate, absent necrosis, and complete surgical excision [16]. In one study of 12 patients with primary pleuropulmonary synovial sarcoma (follow-up 12 months-Syearears, mean 21 months), 5 patients died within 2.5 years from the diagnosis and the local recurrence was seen in 75% of patients within the first 2 years [16]. In 80% of patients, the local recurrence involved either the surgically treated lobe or the ipsilateral residual lobes [5]. Distant metastasis is often hematogenous and rarely lymphatic. Reported distant metastatic sites include the bone, heart, and brain [1,10,17–20]. Rare complications include Pancoast’s syndrome and pulmonary artery pseudoaneurysm because of tumor invasion [3,15].

Surgical excision, often in the form of lobectomy or pneumonectomy, is the most accepted treatment method aiming for complete tumor resection. This is strongly associated with increased survival [21]. Radiotherapy and chemotherapy are often used as adjuvant treatment options [1,5].

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