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CASE REPORT

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Primary thoracic synovial sarcomas: A case report

Nesrine Kallel | **Darine Sakka** | **Asma Zaafouri** | **Rim Khemakhem** | **Ilhem Yangui** | **Sameh Msaad**

Hospital Hedi Chaker, Pneumology Department of Sfax, Sfax, Tunisia

Correspondence
Nesrine Kallel, Hedi Chaker University Hospital Sfax Tunisia, Ain Avenue Km 0,5. 3029 Sfax, Tunisia.
Email: kallel.nesrin@yahoo.com

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Abstract

Primary pulmonary synovial sarcoma (SS) is a rare neoplasm. Its clinicoradiologic attributes are not yet well defined. We report the observation of a patient followed for primary pulmonary synovial sarcoma. We report the radio-clinical features of this rare tumor.

KEYWORDS

chest imaging, chest wall, clinical features, synovial sarcoma

1 | INTRODUCTION

Synovial sarcomas (SS) are high-grade malignancies. They most commonly affect the soft tissues of the extremities, mainly in a periarticular distribution.[1]

Thoracic involvement is rare and includes parietal, pulmonary, and mediastinal sarcomas.[2] Primary thoracic sarcomas and especially lung sarcomas are very rare compared with metastatic sarcomas.[2,3]

Primary pleuro-pulmonary synovial sarcoma (SS) was first described in 1995.[1] The cases described focused on the histopathological features of the tumor. The radiological features of primary thoracic SS have not yet been studied in a formal series. Some authors have attempted to define the radiological characteristics of primary thoracic SS and to correlate the findings with clinical and pathological features.[1]

Through this observation of a patient with a primary synovialosarcoma of the lung, we describe the clinical and radiological features of this rare thoracic tumor.

2 | OBSERVATION

We report the case of a 61-year-old patient, smoking 40 pack-year with a history of ischemic heart disease and hypertension. The patient consulted for a persistent right chest pain evolving for 3 months with low abundance hemoptysis in a context of weight loss of 3–4 kg for 2 months, asthenia, and anorexia. A series of explorations eliminated a cardiac origin of the thoracic pain. Chest X-ray revealed a lung mass measuring about 10 cm [Figure 1]. Chest CT confirmed the presence of a large heterogeneous mass in the right upper lobar, massively necrotic, adhering to the trachea, laminating the superior vena cava, and pushing the esophagus to the left, measuring 14 cm long [Figure 2] without notable mediastinal, hilar, or axillary adenopathy, or adjacent bone involvement. A CT-guided lung biopsy concluded that the pulmonary synovialosarcoma was largely necrotic [Figure 3]. In view of this histological type, a meticulous clinical examination did not find a primary tumor, particularly in the soft tissues and

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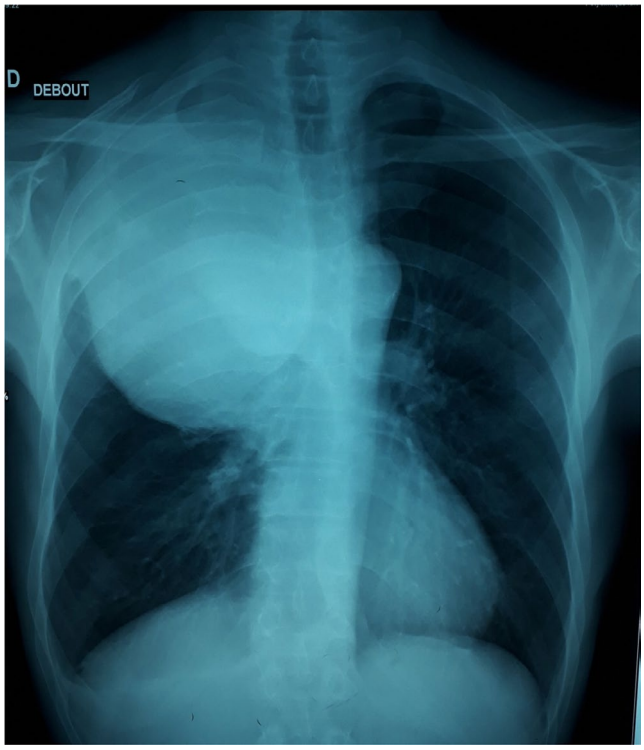


FIGURE 1 Lung mass measuring about 10 cm

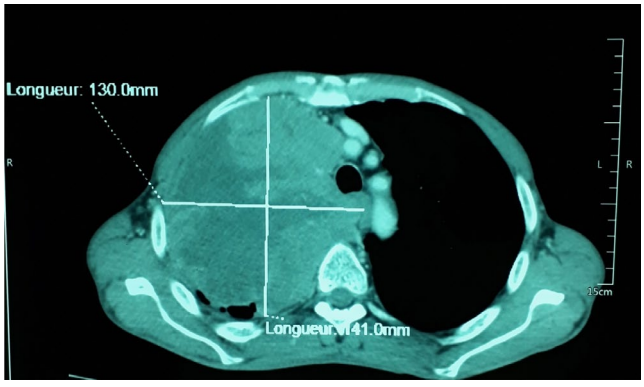


FIGURE 2 A large heterogeneous mass in the right upper lobar, massively necrotic

in the periarticular area; so, the primary pulmonary origin was retained. The remote extension assessment was negative. This case was discussed at a multidisciplinary concertation meeting and the resectability of the tumor was rejected in view of the locoregional extension and mediastinal invasion. The patient was referred to the oncology department for radiochemotherapy.

3 | DISCUSSION

Synovial sarcoma are rare malignant tumor. They are highly aggressive on soft tissue. Their pathogenesis is unknown.[4] They account for 7%–8% of malignancies of mesenchymal origin.[2,5]

Synovial sarcoma is found in 90% of cases in the paraarticular regions and in 10% of cases in various anatomical sites unrelated to synovial tissue.[2,3]

It has become increasingly evident that SS can occur in many other sites, including the head and neck, mediastinum, heart, and esophagus. Only recently, some cases of SS occurring in the lungs and pleura have been reported.[6] Approximately, about 30 cases of primary lung site tumors are described in the literature.[3] The site is preferentially peripheral but a few cases are described in the bronchial tree.[7]

Because of its rarity, SS of the chest wall is difficult to diagnose, its primitive nature is difficult to assert, the search for a primitive extra-thoracic tumor is fundamental before making the diagnosis.[8,9]

Only the absence of extra-pulmonary tumor location in the past, at the time of diagnosis and after 2 years of follow-up will attest to the primitive nature of the lung tumor.[3] In our case, no extra-thoracic signs were retained, allowing us to retain the diagnosis of a primary synovialosarcoma of the lung.

Synovial sarcoma usually occurs between the ages of 20 and 40, but can be seen at any age.[1] In our case, the patient is 61-year-old. In the literature, a slight male predominance has been noted.[2] The sex ratio is 1.5 (8.10).

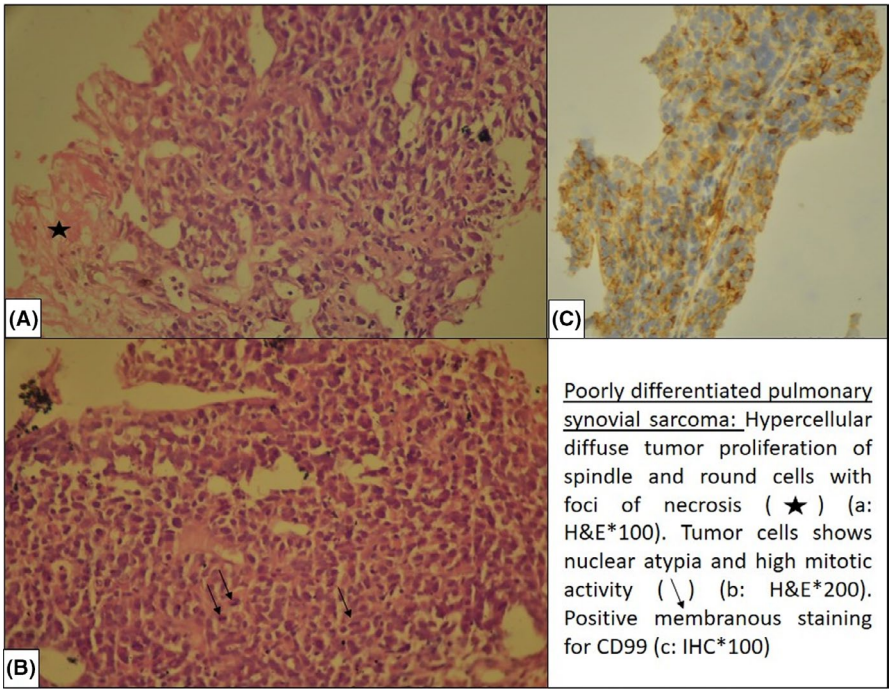
Our clinical case is conformed with the literature results that indicate that the most common symptoms are chest pain, shortness of breath and cough, although up to 24% of patients remain asymptomatic with incidentally identified tumors.[6]

The SS of the chest wall appears as a soft mass, gradually increasing in size over months or years,[1,11] with a vaulted wall as it grows outside. It may be asymptomatic[12] or more often be responsible of a chest pain, gradually increasing, as has been observed in our case over several months.[1] Its intra-thoracic development is most often responsible of dyspnea or cough.[1,11]

A study by A. Duran-Mendicuti et al[1] described the difficulty that can be encountered to assign a specific site to a primary thoracic SS. This study included five patients with SS, the tumor was located in the lungs and/or pleura in one case, in the chest wall in two cases and masses involving both pleuro-pulmonary and chest wall tissue in two cases. In addition, the results highlight the large size (2–10 cm; mean, 6 cm) that can be achieved by a primary thoracic SS.[1]

Chest X-rays are not very effective in determining the characteristics of the tumor, including its size, limits and location in the chest wall, pleura, or lung parenchyma. It reveals calcifications in more than 25% of cases.[8,9] In our case, the chest X-ray showed the presence of an opacity

FIGURE 3 Histopathology aspect



that occupies almost the entire right lung without any image of calcification.

Thoracic CT scan allows a better appreciation of the site of the tumor, its endo- and exo-thoracic extension, and allows to show signs of malignancies such as the heterogeneous appearance with central necrosis, presence of pleurisy, and mediastino-pulmonary invasion.[8]

On thoracic CT scan, the primary SS of the thorax most often presents as a very limited heterogeneous mass without calcifications, without notable mediastinal, hilar, or axillary adenopathy and without bone invasion.[1] Chest wall SS is rarely associated with pleural fluid effusion.[11]

On MRI, about 90% of SS are very limited, sometimes with an encapsulated appearance. The presence of lobulations is frequent.[8]

In their study, Fujimoto et al[1,13] report a chest wall SS showing, on MRI, ovoid and round areas of low signal intensity in T1-weighted images and high signal intensity in.

T2-weighted images within the mass. These authors also described the fluid—fluid levels associated with sedimented blood products as reported in the SS of the extremities. MRI is the best radiological examination to better assess intra-tumoral heterogeneity and locoregional extensions.[10,14]

The diagnosis of SS is based on the anatomopathological, immunohistochemical, and cytogenetic analysis of the tumor tissue.

Macroscopically, it is an oval or rounded tumor, sometimes multinodular, of very variable diameter (0.6–28 cm), often well-defined and encapsulated. It is pale, with white or gray color, with a soft, firm, or rubbery consistency and is dotted with necrotico-hemorrhagic and cystic foci, which reflect the heterogeneous radiological appearance.[7,10,14]

Most tumors have a characteristic t(X; 18) translocation, which involves the SSX1 or SSX2 genes on the X chromosome (Xp11) and the SYT gene on chromosome 18 (18q11). Transcripts of the SYT-SSX fusion gene can be detected on anatomopathological specimens with a sensitivity of 96% and a specificity of 100%.[8] The genetic study was not done in our case.

Surgery remains the main treatment option followed by radiotherapy on the tumor site.[4]

The factors of a good prognosis are as follows: Tumor diameter less than 5 cm, low mitotic index (less than ten mitoses for ten fields at high magnification), low proliferation index (Ki-67 <10%), absence of tumor necrosis, and absence of residual tumor after surgical resection.[2]

Despite the use of new chemotherapy drugs, the prognosis remains poor. Five-year survival ranges from 76% to 35%, and 10-year survival from 63% to 10%.[10]

4 | CONCLUSION

Synovial sarcoma are malignant soft tissue tumors with a rare primary thoracic site. Thoracic SS is manifested by chest pain and dyspnea. Chest CT shows the heterogeneous aspect of the mass. Adenopathy and calcifications are rare. MRI is the gold standard. The diagnosis is anatomopathological. The treatment is essentially surgical as in all soft tissue sarcomas, combining radiotherapy for better local control. However, the prognosis stills poor.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

AUTHOR CONTRIBUTION

This work was carried out in collaboration among all authors. Darine Sakka, Zaafouri Asma, and Khemkhem Rim have made substantial contributions to acquisition and interpretation of data. Kallel Nesrine and Ilhem Yangui involved in drafting the manuscript. Msaad Sameh had given final approval of the version to be published. All authors read and approved the final manuscript.

ETHICAL APPROVAL

Ethics approval has been obtained and preserved by the authors.

CONSENT

Written informed consent was obtained from the family of patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are openly available.

ORCID

Nesrine Kallel https://orcid.org/0000-0002-0622-713X
Rim Khemakhem https://orcid.org/0000-0003-4977-6419

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