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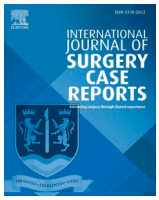

Case report

A case report of primary pleural synovial sarcoma, an uncommon etiology of a thoracic parietal mass

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ABSTRACT

Introduction and importance: Synovial sarcomas are aggressive soft tissue malignancies. Damage to the chest wall is uncommon and rarely seen.

Case presentation: We are reporting an observation of a patient who presented with chest pain and a parietal tumor. Radiological investigations have identified pleural thickening as an extension outside the chest wall. The mass biopsy guided by the scan revealed a synovial sarcoma. The patient underwent a thorough tumor removal surgery with complex postoperative management for hypoxemic pneumopathy.

Clinical discussion: The efficacy of a comprehensive treatment strategy combining surgery, chemotherapy, and radiotherapy for invasive malignancies is still unclear, with total tumor excision being essential. Experts often recommend adjuvant radiation therapy, but the importance of chemotherapy is multifaceted.

Conclusion: We emphasize the rarity of chest localization for these tumors, their unique clinical presentation, and the various therapeutic approaches.

1. Introduction

Synovial sarcoma is a malignant, mesenchymatous tumor that typically arises from soft tissues. It should preferably touch the extremities closest to the major joints [1,2].

Pleural synovial sarcoma is a rare condition; typically, it is associated with a metastatic disease originating from a primary soft tissue tumor. Primary pleural impairment is rare [3–5].

2. Case presentation

A 54-year-old patient, employed in the synthetic fiber industry and a heavy smoker, is experiencing worsening exertional dyspnea and left chest pain during the past 2 months, accompanied by a deterioration in overall health. Seeking medical help.

The examination revealed a mildly uncomfortable 6 cm mass with a soft substance, fixed in relation to the deep plane. We did not find any cases of peripheral adenopathies. The latter phase of the clinical examination emphasizes no particular details.

The chest X-ray revealed an opacity in the left pulmonary hemispheric fields, with blurred boundaries resembling water. A thoracic scan confirmed the presence of pleural thickening near the upper left lobe, in a nodular shape measuring 56 mm × 18 mm, suggesting a pleural tumor [Fig. 1].

The abdominal ultrasound and bronchial fibroscopy both showed normal results. A biopsy, guided by a scan, determined that the mass was synovial sarcoma. An extension test for a primitive tumor came back negative. The surgeon recommended a surgical procedure. A surgical procedure was carried out through the 5th left intercostal space with a horseback incision on the tumor. The surgical procedure revealed a tumor mass infiltrating the parietal pleura, the middle arc of the 3rd, 4th, and 5th left ribs, and the corresponding intercostal spaces.

Moreover, there was no evidence of invasion into the lung tissue. We completely removed the tumor by resecting the infected ribs and reconstructing the chest wall with a propane plate.

Upon examination, the operating room discovered a tumor measuring 6.8 cm in diameter and weighing 250 g. It had a whitish appearance, was mostly cystic, and had firm consistency [Fig. 2]. The surgical unit's anatomopathological examination found a pleural synovial sarcoma that wasn't fully differentiated. It had large round cells, a lot of eosinophilic cytoplasm, and big nuclei with dense chromatins. Dense, diffuse, compact layers of tumor cells surround a significant nerve network and numerous individual striated muscle fibers, all without any signs of necrosis.

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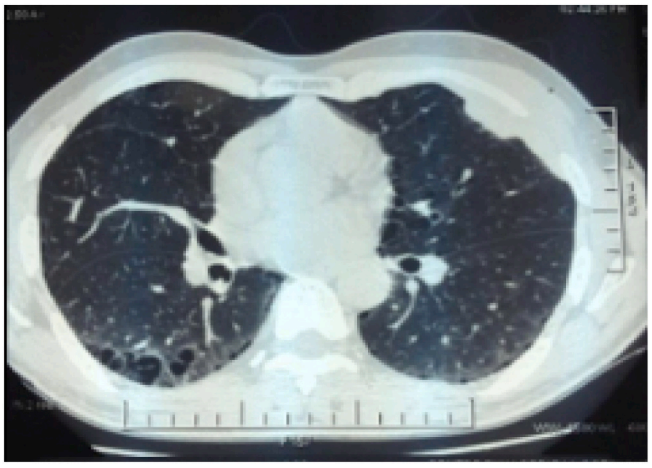

Fig. 1. Localized thickening of the pleura.

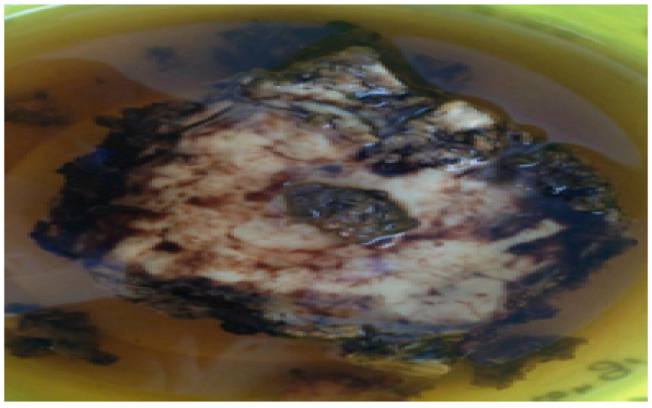

Fig. 2. Macroscopic appearance of the tumor.

After surgery, hypoxemic pneumopathy complicated the patient's recovery, necessitating prolonged mechanical breathing and therapy with broad-spectrum antibiotics. Despite medical intervention, the patient died the 10th day after surgery due to septic shock and a refractory lung condition.

3. Statement

We have reported this case according to the SCARE criteria [6].

4. Discussion

Diagnosing pleural synovial sarcoma can be difficult due to its rarity and similarities to other pleural neoplasms, such as sarcomatous mesothelioma [1]. The clinical presentation includes chest discomfort, shortness of breath, cough, and hemoptysis, as described in the literature [2].

The deep tissue in our patient's thoracic wall was home to a painless tumor. In a thorough investigation by Zeren et al., 40 % of patients were asymptomatic, and a chest X-ray accidentally revealed the diagnosis for a different condition [10]. Common radiological observations include a sizable, varied pleural mass with clearly defined boundaries, an enlarged pleural space, and no indication of mediastinal lymph node involvement [7,11–14].

However, hemothorax and recurrent pneumopathy were the first signs. Calcifications were present in 30 % of cases. There have been cases of mediastinal masses that appear similar to thymomas or germinal tumors [15,16]. Our patient had an atypical growth outside the chest cavity that extended into adjacent ribs.

In these cases, non-specific imaging findings lead to a definitive diagnosis through histological, immunohistochemical, and potentially cytogenetic analysis. Actually, it can be categorized into three distinct anatomopathological forms: the monophasic form and the biphasic form [8,9].

From a macroscopic perspective, biphasic synovial sarcomas consist of two components: cylindrical epithelial cells and fusiform cells. Immunohistochemistry shows that membrane antigen and cytokeratin are expressed by epithelial cells, while vimentin is expressed by fusiform cells. This helps tell synovial sarcoma apart from other malignant pleural tumors, such as mesothelioma [4,17]. The chromosomal translocation t(X;18) (p11;q11), which implicates the SS18 gene and either SSX1, SSX2, or SSX4 genes, induces many tumors.

Translocation affects several oncogenetic pathways, such as the canonical pathway, the SWI/SNF chromatin remodeling complex, and the polycomb repressor complex. The translocation partner has an influence on epithelial differentiation. (SS18-SSX1: 60–70 % monophasic, 30–40 % biphasic) SS18-SSX2 (97 % monophasic, 3 % biphasic) [19].

After confirming the diagnosis of pleural synovial sarcoma, it's crucial to rule out primitive extra-thoracic lesions to prevent the possibility of pleural metastases from a distant synovial sarcoma [19].

The treatment of pleural synovial sarcomas remains challenging due to the rarity of these tumors. Treating pleural synovial sarcomas remains challenging [20]. Although experts have recommended an integrated approach involving surgery, chemotherapy, and radiotherapy, its effectiveness is still uncertain [21–23].

Surgery, especially complete tumor removal, is crucial in treating these invasive tumors. Incomplete resection increases the chances of local relapse and lowers survival rates. It is common to recommend adjuvant radiation therapy after incomplete or extensive resection of large tumors. It is challenging to assess the role of chemotherapy in treating this rare disease, as the effectiveness of treatments containing phosphamide and doxorubicin varies [24]. Neoadjuvant chemotherapy was administered to certain patients to streamline the surgical process by decreasing tumor size and possibly addressing micro metastases [21].

Unfortunately, the outcomes did not meet expectations. Radiofrequency ablation may serve as an alternative for high-risk surgical patients. The following factors contribute to a poor prognosis: the initial tumor size of more than 5 cm in diameter; local invasion; high histological fat; and incomplete surgical resection. Only a complete surgical resection of the tumor remains the determining factor for long-term survival [25].

5. Consent

The patient provided written informed consent for the publication of this case report and its accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

6. Conclusion

Primary pleural synivial sarcoma is a rare tumor. His diagnosis was based on an anatomopathological examination and an immunohistochemical study.

His treatment is primarily surgical, as is the case with all soft-part sarcomas, combining radiation therapy for better local control. However, his prognosis remains reserved.

Author contribution

The report's conception and coordination were undertaken by all authors, who also drafted the manuscript. Furthermore, it should be noted that the final version has been reviewed and approved by all authors.

Ahmed Lasmar: case design.
Wissal Rouabeh: writing the paper.
Imen Mgarrech: literature collection.
Taieb Cherif: literature analysis.
Bechir Ben Radhia: study concept.

Ethical approval

The acquisition and assessment of patient health data was conducted with the patient's written consent. The research was granted approval by the Ethics Committee of Sahloul hospital on 15 March 2023.

Guarantor

Pr Chokri Kortas.

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Conflict of interest statement

There are no conflicts of interest in this report.

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