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Primary Pleural Synovial Sarcoma : A Rare and Agressive Presentation in a 53 – Year - Old Female Patient

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Abstract : Synovial sarcoma is a rare mesenchymal tumor, usually found in the extremities. Its primary occurrence in the pleura is exceptional and presents a major diagnostic and therapeutic challenge. We report the case of a 53-year-old female patient presenting with progressive chest pain and dyspnea. Radiological imaging (chest X-ray and CT scan) revealed multiple large pleural masses with bilateral nodules. Pleural biopsy showed a spindle cell proliferation with mesenchymal features. Immunohistochemical analysis revealed strong expression of TLE1, while cytokeratins AE1/AE3, SATB2, S100, and desmin were negative. This immunophenotypic profile, combined with the morphological features, supported the diagnosis of pleural synovial sarcoma. In the absence of genetic confirmation of the t (X ;18) (p11 ; q11) translocation, the diagnosis relied on strong histopathological and immunohistochemical suspicion. Chemotherapy with anthracyclines was planned, but the patient unfortunately died before treatment initiation.

Pleural synovial sarcoma is an extremely rare entity whose diagnosis depends on morphological, immunohistochemical, and ideally molecular findings. Its clinical presentation is often nonspecific, and its course can be rapidly unfavorable. TLE1 expression is a sensitive marker but not specific, highlighting the importance of a multidisciplinary diagnostic approach. This case underlines the necessity of considering synovial sarcoma in the differential diagnosis of unusual pleural masses, especially in younger patients. Rapid management is essential, although the prognosis remains poor in aggressive forms.

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I. INTRODUCTION

Synovial sarcoma is a rare malignant mesenchymal tumor, accounting for approximately 5-10% of all soft tissue sarcomas [1]. It primarily affects young adults and typically develops in the extremities, especially around large joints. Despite its name, this tumor does not arise from synovial tissue but from undifferentiated mesenchymal cells [2,3].

Primary localization in the pleura is exceptional, with fewer than 50 cases described to date [4]. Such rare pleural tumors pose significant diagnostic challenges, as they may mimic other pleural neoplasms, including sarcomatoid mesothelioma or fibrosarcoma [4,5].

Diagnosis relies on a combination of clinicopathologic criteria: histology, immunohistochemistry—particularly detection of TLE1, a sensitive (82–96%) but nonspecific marker—and ideally, molecular confirmation of the t (X ;18) (p11 ; q11) translocation [6–8]. While TLE1 is strongly expressed in most synovial sarcomas, other mesenchymal tumors may also express it variably [7,8].

We present a dramatic case of pleural synovial sarcoma diagnosed in an adult patient, whose clinical deterioration occurred before chemotherapy could begin. This case highlights the severity of this rare entity and the critical importance of early diagnosis to improve prognosis.

II. CASE PRESENTATION

A 53-year-old female patient with no significant medical history presented with left-sided chest pain and exertional dyspnea. Clinical examination revealed dullness to percussion and decreased breath sounds at the left lung base.

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Chest radiography revealed a large left pleural effusion. Thoracic CT confirmed multiple bilateral pleural nodules and masses, predominantly on the left side, associated with a large heterogeneous left pleural effusion and a smaller right-sided effusion.

The patient underwent pleural drainage followed by video-assisted thoracoscopic surgery (VATS) for diagnostic purposes. Direct visualization during VATS showed multiple pleural masses, and multiple biopsies were obtained.

Histological analysis revealed a dense spindle cell proliferation with low-grade atypia, arranged in intersecting fascicles. Immunohistochemistry showed strong expression of TLE1 and negativity for AE1/AE3, SATB2, S100, and desmin. These findings supported the diagnosis of monophasic spindle cell pleural synovial sarcoma.

Genetic confirmation of the t (X ;18) (p11 ; q11) translocation was not available. However, the morphologic and immunohistochemical features were strongly suggestive of the diagnosis.

A fluorodeoxyglucose positron emission tomography (18-FDG PET-CT) scan was performed as part of the staging workup. It showed a right-sided hypermetabolic pleural process (SUV max : 5.6) associated with a pathological hypermetabolic lymph node in station 3a (SUV max : 3.9).

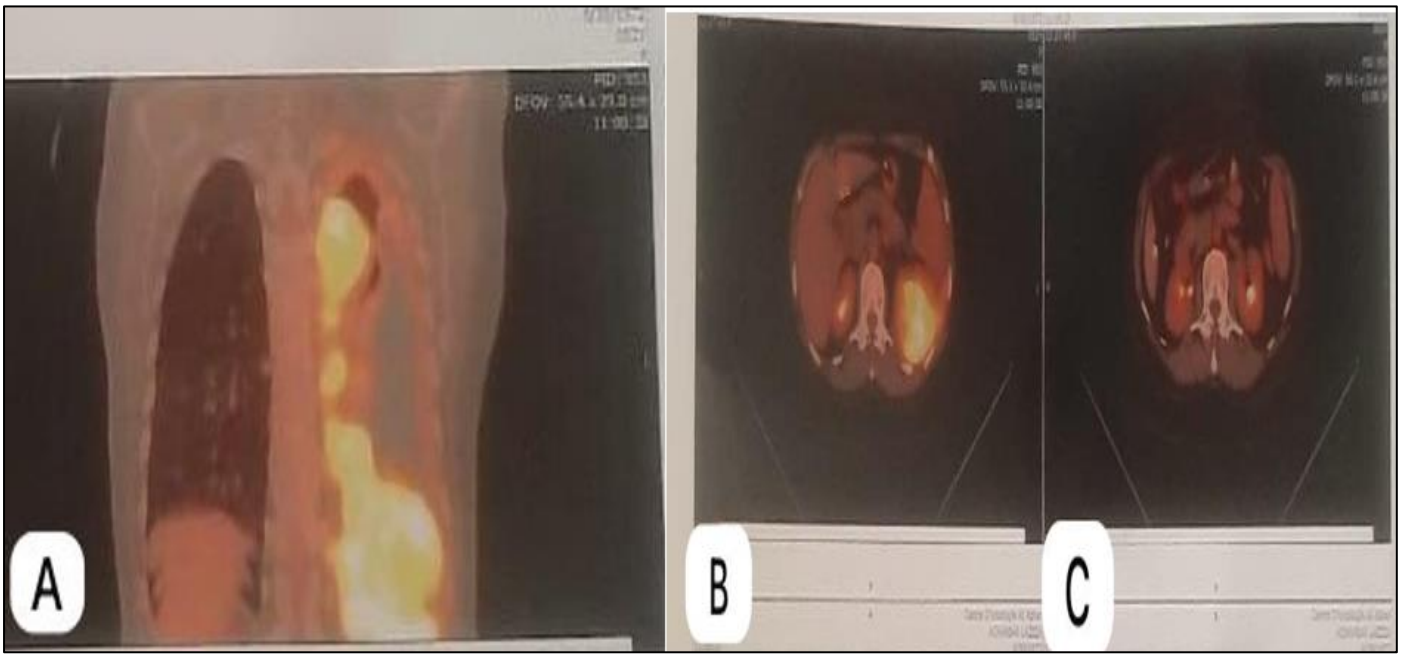

Fig 1 A : Intense FDG Uptake in the Left Hemithorax, Corresponding to the Postero-Lateral Pleura. B : Well-Localized FDG Uptake at the Postero-Lateral Aspect of the Left Pleura. C : No Suspicious Hypermetabolic Activity in the abdominal Organs, Adrenal glands, or Kidneys.

III. DISCUSSION

Primary pleural synovial sarcoma (PPSS) is an extremely rare tumor, with often nonspecific clinical presentations. The most common symptoms include chest pain, cough, dyspnea, or pleural effusion—as seen in our patient. These findings are consistent with most reported cases, such as those described by Bégueret et al. [9] and Dennison et al. [10].

Radiologically, thoracic imaging may reveal unilateral or bilateral pleural masses, sometimes associated with effusions. However, these findings are not specific and may mimic other entities such as mesothelioma or pleural metastases, necessitating early histological sampling [11].

Histologically, the monophasic spindle cell variant is the most frequent in PPSS. It can mimic other tumors, notably sarcomatoid mesothelioma or fibrosarcoma. Therefore, immunohistochemistry is essential for differential diagnosis. As noted by Foo et al. [12], strong TLE1 expression combined with negative cytokeratin, S100, and desmin supports the diagnosis of synovial sarcoma.

However, TLE1 is not entirely specific and can be expressed in other sarcomas. This makes detection of the t(X;18)(p11;q11) translocation the most reliable confirmation method. In resource-limited settings, where molecular testing is not always accessible, diagnosis often relies on combined morphological and immunohistochemical features, as reported by Amodio et al. [13].

The standard treatment is complete surgical resection when feasible. In locally advanced or unresectable cases, anthracycline-based chemotherapy is commonly employed. Nonetheless, the response remains modest. According to studies by Issels et al. [14] and Palmerini et al. [15], overall survival in advanced cases treated with chemotherapy ranges between 12 and 36 months.

In our case, the patient died before chemotherapy could be initiated, underscoring the potentially fulminant course of

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this tumor. This case highlights the importance of early diagnostic suspicion, rapid thoracoscopic evaluation, and a coordinated multidisciplinary approach.

IV. CONCLUSION

Primary pleural synovial sarcoma (PPSS) is a rare and aggressive tumor, often diagnosed late due to its nonspecific clinical presentation. Our case illustrates the diagnostic and therapeutic challenges associated with this entity.

A combination of thorough histological examination, targeted immunohistochemistry (TLE1), and, when possible, molecular confirmation is essential for accurate diagnosis. The absence of t (X ;18) translocation does not rule out the diagnosis when other features are strongly suggestive.

Management ideally involves complete surgical resection. When surgery is not possible, chemotherapy remains the main alternative, although its efficacy is limited. In all cases, a rapid multidisciplinary approach is critical.

This case emphasizes the importance of considering PPSS in the differential diagnosis of unusual pleural masses and highlights the need to improve diagnostic tools in low-resource settings to optimize care for these rare but severe tumors.

The authors have declared that no competing interests exist.

HUMAN ETHICS

Consent was obtained or waived by all participants in this study.

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