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Primary Mediastinal Synovial Sarcoma; A rare case

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ABSTRACT

Synovial sarcoma, a rare subtype of soft-tissue sarcomas, typically manifests in the extremities of young individuals. Mediastinal occurrences are exceedingly uncommon, with sparse cases documented. This article details the atypical presentation of a mediastinal synovial sarcoma in a 68-year-old female patient, diverging from the common demographic and anatomical predilections. Initial symptoms of a dry cough and fatigue led to the discovery of a mediastinal mass via chest CT scan. Subsequent pathological examination confirmed the diagnosis of mediastinal synovial sarcoma. Despite the identification of this rare tumor, the patient's condition progressed rapidly, resulting in her demise within two months post-diagnosis. This case underlines the critical need for increased awareness and research into the mediastinal variant to improve diagnostic accuracy and treatment efficacy. Despite synovial sarcoma being predominantly identified in the extremities, occurrences in the mediastinum present unique diagnostic dilemmas and therapeutic hurdles due to limited literature and atypical clinical presentations. The utilization of advanced imaging modalities, immunostaining techniques, and multimodal treatment approaches is essential in the management of mediastinal synovial sarcoma.

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Introduction

Synovial sarcoma is the fourth most common type of soft-tissue sarcoma, representing 2.5%–10.5% of primary soft-tissue malignancies globally. It predominantly affects the extremities (80%–95% of cases), especially the knee in the popliteal fossa, of adolescents and young adults aged 15–40 (1). However ,a primary occurrence in the mediastinum is quite rare with only a few reported cases in the world literature (2). While synovial sarcoma in the extremities has well-established diagnostic criteria and treatment protocols, the mediastinal variant poses unique diagnostic and therapeutic challenges due to its atypical presentation and limited literature.

In our case, we present a 68-year-old woman presented with a 2-month history of dry cough and fatigue underwent a chest CT scan, revealing a mass in the mediastinum. The pathological result confirmed a rare tumor known as synovial sarcoma of the mediastinum. Unfortunately, she passed away two months after the diagnosis.

Case Report

A 68-year-old female, sought medical attention due to persistent symptoms of dry cough and generalized fatigue that had been troubling her for the past two months. She underwent a comprehensive evaluation that included a spiral chest HRCT scan to investigate the underlying cause of her symptoms.The results of the chest CT scan revealed the presence of a suspicious mass located within the mediastinum, prompting further investigations to characterize the nature of the lesion. The CT scan showed an oval-shaped

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fluid density mass with sharp margin< measuring about 138*95mm located in superior and posterior mediastinum extending to middle mediastinum associated with adjacent atelectasis in left lung and right upper lobe. The mentioned mass did not show calcification, bony destruction or spinal canal extension, but encased aortic arch and pushed trachea, main bronchi and esophagus anteriorly. (figures 1&2) The mass that was not present in the chest HRCT scan performed for the patient two years ago. After obtaining a percutaneous CT-guided core biopsy from the mass, the report was unexpectedly surprising. The histopathological analysis of the ct-guided core biopsy from the mass revealed an infiltrative neoplastic growth characterized by fascicles of spindle cells with elongated hyperchromatic nuclei and evidence of mitotic activity. These microscopic features are indicative of a malignant process, warranting further immunohistochemical staining for a

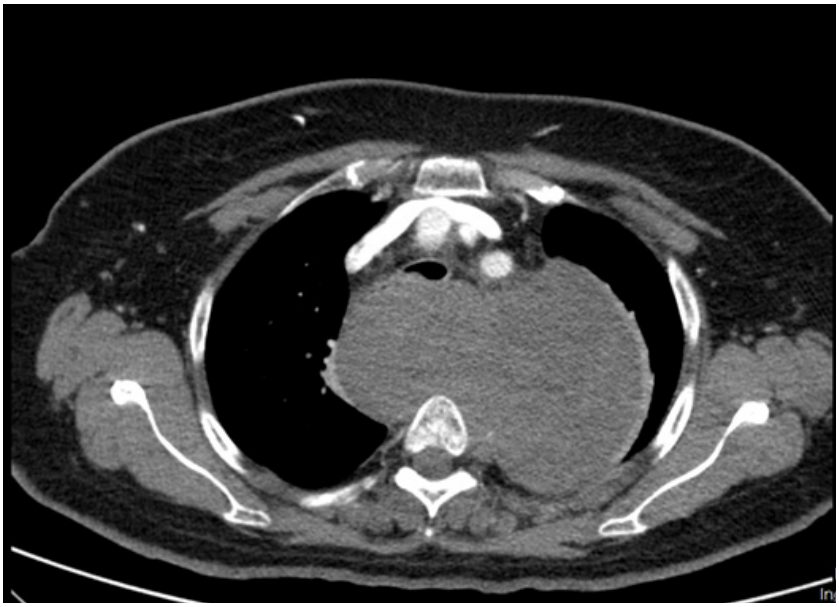

Figure 1. Axial spiral chest CT scan with IV contrast showed an oval-shaped fluid density mass with sharp margin measuring about 138*95mm located in superior and posterior mediastinum extending to middle mediastinum associated with adjacent atelectasis in left lung and right upper lobe. The mentioned mass did not show calcification, bony destruction or spinal canal extension, but encased aortic arch and pushed trachea, main bronchi and esophagus anteriorly

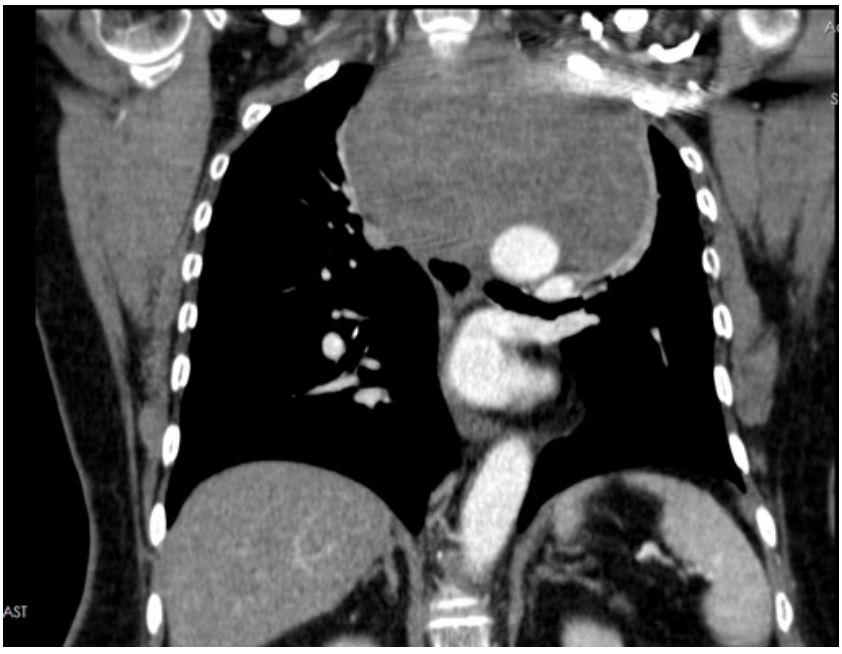

Figure 2. Coronal view of spiral chest CT scan with IV contrast

definitive diagnosis.

The subsequent immunohistochemistry (IHC) evaluation yielded a distinct pattern of marker expression (Table 1). Negative results were observed for CK, S100, SMA, Desmin, CD34, SOX10, H.Caldesmon, WT1, and Calretinin, indicating the absence of specific markers associated with different cellular lineages or tissues. Notably, the Ki67 marker showed proliferative activity in 10% of tumor cell nuclei, suggesting a moderate level of cellular proliferation.

The diffusely and strongly positive expression of TLE1 in the tumor cells is a hallmark finding in the IHC panel. TLE1 positivity is a sensitive feature for synovial sarcoma and is considered a highly sensitive and specific marker for this entity (3). The overall IHC profile, in conjunction with the histologic features, led to the interpretation of Monophasic Spindle cell Synovial Sarcoma.

This comprehensive IHC analysis aligns with the established diagnostic criteria for Monophasic Synovial Sarcoma, reaffirming the significance of immunohistochemical profiling in refining the diagnosis of rare soft tissue malignancies. The identification of TLE1 positivity further solidifies the classification of the tumor, emphasizing the importance of employing a multidisciplinary approach and advanced diagnostic techniques in the accurate characterization and classification of challenging neoplastic entities like synovial sarcoma.

Despite the efforts of the medical team to diagnose and treat her illness, the lady was not satisfied with the treatment and did not undergo the surgery. She did not accept the medical interventions and unfortunately passed away approximately two months later. However, through this article, we hope that she can also help other rare patients by sharing her disease story.

Discussion

Approximately 10-20% of cases of Synovial sarcoma are found in areas outside of the extremities (1). In general, Soft tissue sarcomas, which include angiosarcoma, leiomyosarcoma, sarcomatoidmesothelioma, rhabdomyosarcoma, and Synovial sarcoma, make up less than 0.01% of all malignant thoracic neoplasms (4). There are two types of synovial sarcoma, monophasic and biphasic. The better recognized variant is the biphasic synovial sarcoma which consists of proliferation of bland looking spindle-shaped cells in a collagenous background and hemangiopericytomatous vascular growth pattern. Monophasic tumors can show either spindle cells or epithelial components (5). Immunostains have proven to be invaluable in confirming the diagnosis of synovial sarcoma. The presence of epithelial markers in both the gland-like and spindle cell components lends support to the diagnosis. Among these markers, EMA is frequently found to be positive (6). Furthermore, pan-cytokeratin, cytokeratin 7, and cytokeratin 19 may also exhibit positivity in both the epithelial-like and spindle cell components (7). Some cases may show positivity for bcl-2 as well (8). More recently, TLE1 has emerged as a promising diagnostic marker for synovial sarcoma, purportedly offering higher sensitivity and specificity compared to other markers (9).

Accurate staging of the disease is crucial for

Table 1. Immunohistochemical Study Report

Markers	Description of Reaction
CK	Negative
S100	Negative
SMA	Negative
Desmin	Negative
CD34	Negative
SOX100	Negative
H.Caldesmon	Negative
WT1	Negative
Calretinin	Negative
Ki67	Proliferative activity in 10% of tumor cells
TLE1	Diffusely and strongly positive in tumor cells

effective patient management, necessitating assessment of both the primary tumor and distant disease. While magnetic resonance imaging (MRI) has conventionally been utilized for evaluating soft tissue masses, Positron Emission Tomography (PET) has emerged as a valuable tool due to increased 18F-FDG uptake observed in soft tissue sarcomas (10). The optimal therapeutic strategy for individuals diagnosed with mediastinal synovial sarcoma entails complete surgical excision, which stands as the sole factor correlated with enhanced survival outcomes. In cases where the disease is unresectable but non-metastatic, neoadjuvant chemotherapy and radiotherapy should be deliberated prior to surgical intervention. For patients deemed ineligible for surgical resection, the administration of chemotherapy, particularly high-dose ifosfamide with or without doxorubicin, may be warranted (11). The incorporation of adjuvant chemotherapy and radiotherapy into a comprehensive multimodal treatment regimen is recommended for all patients. Despite the availability of these multifaceted treatment modalities, the prognosis for individuals with mediastinal synovial sarcomas remains grim, with a median overall survival of 36 months and a 5-year overall survival rate of 35.7%, contrasting with a 5-year overall survival rate of 50–80% observed in cases of extremity primaries (11). This disparity in outcomes is likely attributed to factors such as the advanced stage of tumor presentation, substantial tumor size, challenges in achieving complete surgical resection due to involvement of critical anatomical structures, and the high incidence of the poorly differentiated subtype. Continued research, case documentation, and collaborative efforts will be essential in unraveling the underlying mechanisms, and clinical implications of these rare conditions to provide optimal care and outcomes for affected individuals.

In conclusion, the case of mediastinal synovial sarcoma presented in this article sheds light on the diagnostic challenges, therapeutic complexities, and grim prognosis associated with this rare malignancy. Despite synovial sarcoma being predominantly identified in the extremities, occurrences in the mediastinum present unique diagnostic dilemmas and therapeutic hurdles due to limited literature and atypical clinical presentations. The utilization of advanced imaging modalities, immunostaining techniques, and multimodal treatment approaches is essential in the management of mediastinal synovial sarcoma. While surgical resection remains the cornerstone of treatment, the incorporation of neoadjuvant chemotherapy, radiotherapy, and adjuvant therapies may offer additional benefits in certain cases.

By elucidating the intricate nature of mediastinal synovial sarcoma and exploring personalized treatment strategies, we strive towards better patient care and management of this challenging malignancy. This case serves as a poignant reminder of the importance of vigilance in recognizing atypical presentations of soft tissue sarcomas, the significance of early intervention, and the crucial role of interdisciplinary collaboration in combating rare and aggressive malignancies such as mediastinal synovial sarcoma.

Acknowledgements

This case report was obtained from a patient in Rafsanjan Hospital, and the pathological results of this case was confirmed at the Pathology Laboratory of DEGHAT in Tehran.

Conflict of interest

The authors declare that they have no competeing interest.

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