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Primary Mediastinal Synovial Sarcoma: A Rare Case Report

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Abstract

Background: Synovial sarcomas commonly occur in the extremities of young adults. A primary occurrence in the mediastinum is very rare with only a few reported cases in the world literature.

Case Report: This paper is about a 47-year-old male who presented with retrosternal chest pain and shortness of breath on exertion. Imaging showed an anterior mediastinal mass. Pathological examination of the resected mass showed a biphasic neoplasm with a spindle cell component admixed with gland-like elements. The tumor showed positive staining with cytokeratin, epithelial membrane antigen, and vimentin confirming the diagnosis of a biphasic synovial sarcoma.

Conclusions: A wide range of neoplasms, both primary and metastatic, occurs in the mediastinum, which poses considerable diagnostic difficulties. A synovial sarcoma should always be considered in the differential diagnosis, and immunohistochemistry is an important adjuvant tool in this situation. This paper highlights the importance of recognizing an unusual presentation of this aggressive neoplasm to aid appropriate clinical management.

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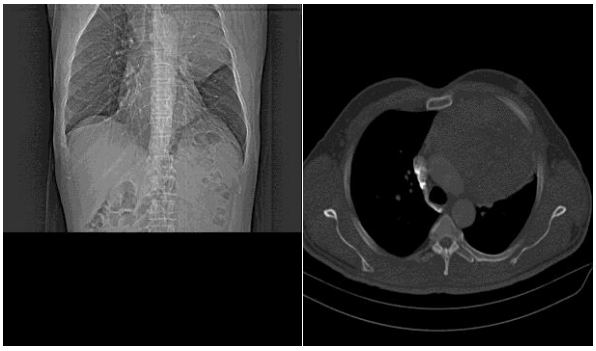
Introduction

Synovial sarcoma is a mesenchymal tissue cell tumor that exhibits epithelial differentiation. Most frequently, it arises in the extremities of adolescents and young adults (1), while a primary occurrence in the mediastinum is quite rare with only a few reported cases in the world literature (2). Primary mediastinal synovial sarcoma is a malignant tumor, able to invade adjacent organs or give distant metastases. Pathological examination is crucial to establish the diagnosis, whereas clinical presentation and imaging patterns are often aspecific and misleading. Herein, we report a case of 14 cm primary mediastinal synovial sarcoma, diagnosed in a 47-year-old male.

Case Report

A 47-year-old man presented with symptoms of retrosternal chest pain, not related to physical activity or exertion. He also complained of shortness of breath on exertion. There was no history of cough, hemoptysis, fever, or weight loss. Physical examination did not reveal anything significant. His blood investigations at presentation revealed hemoglobin level of 12.5 g/dl, total leukocyte counts of 8500 cells/mm^3, and platelet counts of 360,000/mm^3. His serum electrolytes, renal function tests, and liver function tests were within normal range. Germ cell tumor markers such as lactate-dehydrogenase, alpha-fetoprotein, and β-human chorionic gonadotropin levels were within normal limits. The chest X-ray showed a large mass in left thoracic cavity, adjacent to the mediastinum (Figure 1a). Computed tomography (CT) scan revealed a 14 cm heterogeneously anterior mediastinal mass (Figure 1b).



a **b**

Figure 1. (a) Chest X-ray showed a large mass in left thoracic cavity, (b) computed tomography scan of the chest revealed a 14 cm heterogeneously anterior mediastinal mass

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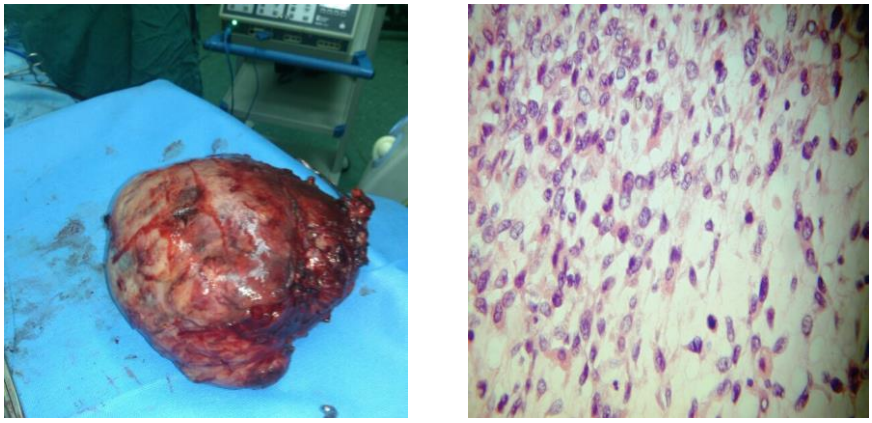

a **b**

Figure 2. (a) Tumor removed at sternotomy, (b) histology demonstrating spindle and epithelioid cells

CT guided biopsy was done. It was reported as sarcoma. Positron emission tomography (PET) scan was performed to exclude any metastatic disease (no other foci that may represent metastatic disease). Then, the patient underwent median sternotomy and excision of tumor was performed. The tumor measured about 14 cm × 14 cm × 9.5 cm (Figure 2a). Histologically the tumor was composed of spindle cells and glandular epithelial structures (Figure 2b). Numerous mitotic figures (more than 4/10 high-power field) were seen, and there were focal areas of necrosis. Immunohistochemistry demonstrated strong positivity for cytokeratins, vimentin and focal positivity for epithelial membrane antigen (EMA). Mesothelial markers and primary pulmonary tumor marker thyroid transcription factor-1 (TTF-1) were negative. With the combination of histopathological features and immunohistochemical findings, a diagnosis of a primary biphasic synovial sarcoma of the mediastinum was offered. Post-operative recovery was smooth. The patient received adjuvant chemotherapy. The patient is on regular follow-up without evidence of residual or recurrent disease.

Discussion

The term synovial sarcoma is a misnomer because the tumor does not arise from the synovium; it only resembles synovial tissue on light microscopy. It appears to arise from multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures and lack synovial differentiation (3,4). Less than 10-20% of synovial sarcoma arise in extra-extremity locations (1,5). In general, soft tissue sarcomas including angiosarcoma, leiomyosarcoma, sarcomatoid mesothelioma, rhabdomyosarcoma, and synovial sarcoma, account for < 0.01% of all malignant thoracic neoplasms (6). According to data from a large population-based study, around 17% of new cases of soft tissue sarcomas (including variety of histologic subtypes) arise in thoracic locations including the pleura, lungs, and the mediastinum with an approximate incidence of 6/million populations (7). Nevertheless, there are no data that accurately report the exact incidence of synovial sarcoma cases that arise primarily in the mediastinum. Morphologically, synovial sarcomas are divided into monophasic and biphasic subtypes. The biphasic variant consists of proliferation of bland looking spindle-shaped cells, along with evidence of epithelial differentiation. Monophasic subtypes on the other hand can show either spindle cells only or occasionally epithelial component only (8). A poorly differentiated variant of synovial sarcoma is also recognized (9). The differential diagnosis of a primary synovial sarcoma in the mediastinum is complex as a wide array of primary and metastatic tumors occur in this site. Immunohistochemistry can be very helpful. Immunostains are valuable and supportive for the diagnosis of a suspected case of synovial sarcoma. The expression of epithelial markers in the gland-like component and more importantly in the spindle cell component supports the diagnosis. EMA is the most commonly positive marker among all epithelial markers (9). Pan-cytokeratin and cytokeratins 7 and 19 can also be positive in the epithelial-like component as well as the spindle cell component (10). In addition, positivity for bcl-2 can be seen in some cases (11).

Accurate staging of the disease is important for appropriate patient management and requires the evaluation of the primary tumor and assessment for distant disease. Magnetic resonance imaging has been the traditional imaging tool for assessment of soft tissue masses, but more recently soft tissue sarcomas are well recognized as showing increased 18F-fluoro-2-deoxy-d-glucose uptake in PET (12). Broad surgical resection is the cornerstone of therapy. Complete resection of the tumor was the overwhelming factor in determining survival in a review of primary mediastinal sarcomas (13). Radiotherapy is recommended with positive margins (14). The place for chemotherapy of this tumor is not well defined, and in none of the other case, reports were pre-operative

chemotherapy given (15). Combination of adriamycine and ifosfamide has been used as an adjuvant therapy and for recurrences (15). Traditionally, synovial sarcoma has been considered to have a poor prognosis with metastasis occurring in 50% of the cases. The lung is the most common site for metastasis followed by lymph nodes and bone. However, not all synovial sarcomas share the same dismal outcome anymore. Spillane et al. demonstrated that an age > 20 years at diagnosis and size > 5 cm were associated with a significantly worse prognosis (16,17). However, prognosis after aggressive multimodal therapy is good and survival up to 14 years has been documented (15).

As conclusion, the mediastinum is an uncommon site of occurrence of synovial sarcoma that occurred in index case. Synovial sarcoma must be entertained in the differential diagnosis of spindle cell as well as biphasic tumors arising in the mediastinum. This case report emphasizes the importance of prompt clinical suspicion, accurate histopathological diagnosis, and use of appropriate immunohistochemical markers in the diagnosis of this unusual tumor in an unusual site. Aggressive multimodal therapy is recommended, and our patient has responded to surgery and chemotherapy.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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