CASE REPORT

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Primary Pulmonary Synovial Sarcoma with Hemothorax: a Case Report

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ABSTRACT

Background: Synovial sarcoma is a rare and aggressive soft tissue malignancy most commonly arises from periarticular tissue of the extremities. Although several cases in the literature have reported different origins, primary pulmonary synovial sarcoma (PPSS) is an exceedingly rare and underrecognized entity, accounting for 0.5% of all lung malignancies. Clinical presentation includes chest pain, dyspnea, cough, and hemoptysis. The finding of hemothorax is a rare presentation and was barely reported in the literature. Due to its rarity and aggressive nature, the optimal treatment is unclear, while the mainstay remains surgical resection with chemo- and/or radiation therapy. **Objective:** To report a case of hemorrhagic effusion subsequently diagnosed with primary pulmonary synovial sarcoma with the main objective of enriching the literature regarding this rare malignancy. **Case report:** A 52-year-old male smoker with a background of coronary artery disease, hypertension, and diabetes mellitus was referred to our hospital. The patient presented with a history of chest pain, dyspnea, and massive right-sided pleural effusion. Laboratory investigations were unremarkable except for anemia. Chest x-ray showed a complete opacity on the right lower zone with right-sided pleural effusion. Thoracentesis was done and revealed hemorrhagic exudative effusion. Computed tomography (CT) scan showed a right heterogeneous lung mass compressing the medial segment of the middle lobe. Subsequently, the patient underwent bronchoscopy, which showed compression and edema on the right middle lobe bronchus with traces of blood coming from the right lower lobe. The patient underwent a right posterolateral thoracotomy, a fungating mass eroding the medial segment of the middle lobe was resected that was diagnosed as high-grade primary pulmonary synovial sarcoma. Radiotherapy was instituted. The patient died after two years due to recurrence. **Conclusion:** PPSS is an aggressive disease with poor prognostic outcomes, and Its presentation is almost similar to other lung malignancies. Meanwhile, there is no definitive management guideline, and most management depends on surgical resection if feasible with adjuvant chemo-radiation therapy.

Keywords: Primary pulmonary synovial sarcoma, lung sarcoma, hemothorax, lung cancer, care report.

1. BACKGROUND

Synovial sarcomas often manifest as primary periarticular soft tissue neoplasms, but recent literature has shown that they can also develop from a variety of different sites. Primary pulmonary synovial sarcoma (PPSS) is a rare form of sarcoma that resembles 0.5% of all lung cancers (1). Although the exact source of histogenesis is unknown, immature mesenchymal cells are thought to be the culprit (2). We report a case of a patient who had hemothorax on presentation which is a rare manifestation in a rare entity of a disease.

2. OBJECTIVE

The aim of this article was to report a case of hemorrhagic effusion subsequently diagnosed with primary pulmonary synovial sarcoma with the main objective of enriching the literature regarding this rare malignancy.

3. CASE REPORT

A 52-year-old male, smoker (30 packs/year), with a background of coronary artery disease, hypertension, and diabetes mellitus. The patient was referred to our hospital with a history of chest pain, dyspnea, and massive right-side pleural effusion. The laboratory investigation was unremarkable except for anemia. The chest x-ray showed complete opacity on the right lower zone

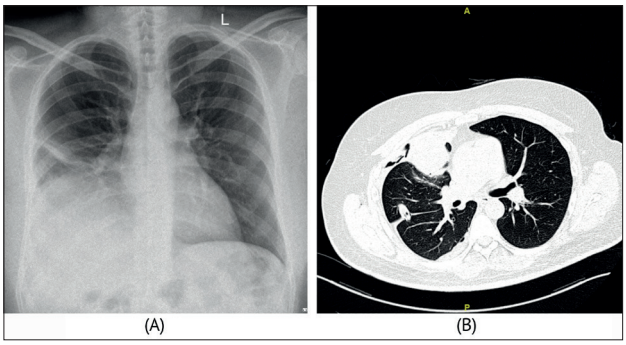

Figure 1. (A) Chest x-ray showing a massive right-side pleural effusion. (B) Computed tomography (CT) scan shows a heterogeneous lung mass noted at the anterior segment of the right lower lung measuring around 8.8 x 7.5 x 7 cm with a right massive pleural effusion involving the superior mediastinal pleura associated with hyperdense nodular pleural thickening causing consolidation of the adjacent lung lobe mainly at the lower lung, it is causing a mass effect upon the mediastinum and cardiac structure with shining toward the contralateral side.

of the chest with right-sided pleural effusion (Figure 1-A). Thoracentesis was done and revealed hemorrhagic exudative effusion. The acid-fast bacilli stain was negative. Subsequently, the patient underwent bronchoscopy which showed compression and edema on the right middle lobe bronchus with traces of blood coming from the right lower lobe.

A chest tube was inserted and drained initially around 1 liter of bloody effusion followed by around 500-750 ml of the same nature each day. Computed tomography (CT) scan showed a right heterogeneous lung mass compressing the medial segment of the middle lobe (Figure 1-B). Furthermore, tumor markers were all within the normal range, including carbohydrate antigen 19-9, carcinoembryonic antigen, and alpha-fetoprotein.

The patient underwent a right posterolateral thoracotomy. There was a fungating mass eroding the medial segment of the middle lobe which was resected. However, there was massive bleeding coming from the base of the mass due to the erosion from the pulmonary vessels, hence middle lobectomy was performed.

A gross histopathological examination of the specimen showed a right lobectomy specimen comprising a triangular piece of lung tissue measuring 9 x 8 x 1 cm along with separate fragments of white to gray-tan, soft to firm tissue, measuring in

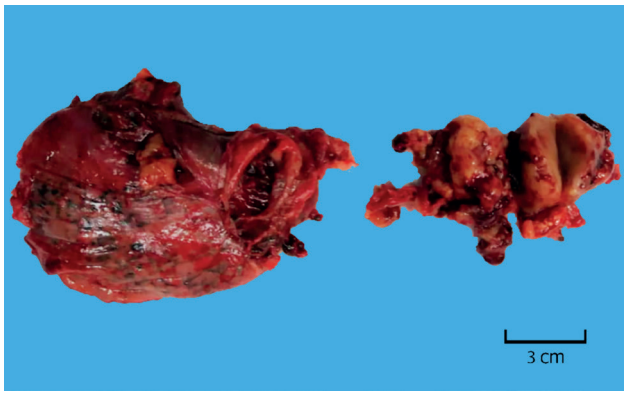

Figure 2. The left structure illustrates the right lung's middle lobe, while the right structure highlights the tumor.

aggregate 8.5 x 5 x 1 cm, the largest measuring 5 x 4 x3 cm (Figure 2). Serial sectioning of the lung tissue was unremarkable while the separate fragments revealed a white to gray tan cut surface. Fragments of right parietal and diaphragmatic pleura measuring 12 x 10.5 x 2 cm and 4.5 x 2 x 0.6 cm respectively and 6 hilar lymph nodes from stations II, IV, and VII were also sampled.

The microscopic examination revealed monophasic neoplasm comprising sheets and fascicles of spindly to ovoid cells with focal hemangiopericytoma like vascular pattern and focal necrosis. The cells revealed stippled chromatin, scattered mitoses, frequently atypical and

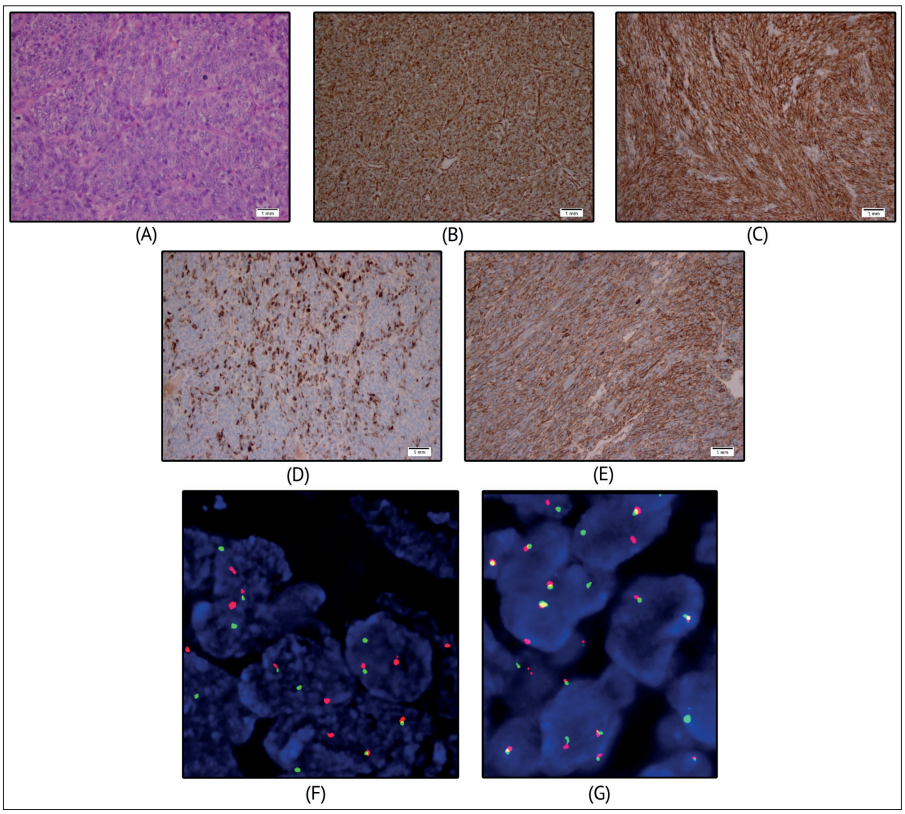

Figure 3. (A) Microscopic examination shows a monophasic spindle cell neoplasm with cells arranged predominantly in sheets and fascicles with foci of hyalinization and lobulation. The cells revealed pleomorphism. The nuclei were irregular, with stippled chromatin, prominent nucleoli and scattered mitosis, frequently atypical (Haemotoxylin and Eosin stain ×40). For (B), (C), (D) and (E) Immunocytochemical analysis of the tumor showing co-positivity for mesenchymal, muscle and epithelial markers as following: Vimentin, CD-56, CK-7 and H-Caldesmon, respectively. (F) and (G) Cytogenetic testing by FISH analysis revealed the tumor cells to be positive for SS18 gene rearrangement.

nuclear overlapping (Figure 3-A). The immunohistochemical analysis of the tumor cells showed positivity for Vimentin, H-Caldesmon, CK-7, CD-99, CD-56, and BCL-2 (Figures 3-B, C, D and E). There was focal positivity for EMA. The Ki-67 proliferation index was high, approximately 90%. No immunoreactivity was seen for Pan-CK, CK-20, Napsin, TTF-1, SMA, S-100, Synaptophysin, Chromogranin, and CD-34 (Figures 3-B, C, D and E). Cytogenetic testing by FISH analysis revealed the tumor cells to be positive for SS18 gene rearrangement (Figure 3-F, and G). The final report confirmed high grade primary pulmonary synovial sarcoma. The right middle lobe, parietal and diaphragmatic pleura and were free of the malignant neoplasm. No metastatic deposits were seen in the lymph nodes. FISH analysis in cytogenetic testing identified a positive SS18 gene rearrangement in the tumor cells (Figure 3 - F and G)

The patient was referred to King Fahad Specialist Hospital in Dammam (KFSH-D) and two phases of radiotherapy were given. The volumetric modulated arc therapy photon type technique was used, (50 Gy/25) at the operation bed and another dose of (10/Gy5).

Two months after the operation, the patient developed a left middle cerebral artery acute infarction secondary to intracranial atherosclerotic disease and ended with a major disability. After 2 months CT chest-abdomen and pelvis (CAP) was done and showed necrotic right pulmonary and pleural-based nodules and masses extending into the right hilum and inseparable from the pericardium, with underlying rib destruction and left upper lobe nodule likely representing metastatic. After 14 months, a positron emission tomography (PET) scan revealed multiple small to large densities seen in the right lung field with SUVmax 2.0. Five months later, there was interval progression of previously noted lung nodules on the right side with newly developed necrotic right hilar lymph nodes. As the patient's condition was poor, the patient was started on comfort care and succumbed to his disease after twenty sixth months after his initial diagnosis.

4. DISCUSSION

The semi-epithelial malignant mesenchymal tumor known as synovial sarcoma (SS) was first believed to have its origins in the synovium (3,4). New research suggests that a pluripotent mesenchymal stem cell may be the genesis of SS, even though its etiology is uncertain (5). PPSS comprises 14.7-18% of all primary lung sarcomas and constitutes around 0.5% of all lung malignancies (5,6). Despite it is rareness most synovial sarcoma is present in extremities that favor metastasizing to the lung forming secondary pulmonary synovial sarcomas (7,8). Synovial sarcoma (SS) is more common in adolescents and young adults (8).

Pulmonary symptoms such as chest pain, dyspnea, cough, and hemoptysis are the frequent manifestation of PPSS (9). The radiological study may reveal ipsilateral hemothorax in a chest x-ray. Hemothorax is likely to be due to lung infiltration by the tumor and erosion of pulmonary vessels and was reported in only three cases (9–11). Upon our knowledge, this is the fourth case of PPSS presenting with hemothorax in the literature. The PPSS appears on CT scan as sharply demarcated lesions without calcification and a heterogeneous appearance with focal necrosis, and hemorrhage also may be encountered (2).

The histological classification of SS is the monophasic spindle, monophasic epithelial, and biphasic (9). The biphasic subtype is poorly differentiated, and it contains both spindle and epithelial patterns (9). The monophasic spindle is the most common subtype among the others (2). The immunohistochemical staining expresses vimentin, cytokeratin, and epithelial membrane antigen positively in SS, while negativity for S100 protein (10). Moreover, the immunoreactivity of the BCL-2 protein distinguishes synovial sarcoma from other possibilities (10). Significantly, our case was positive for CD-99 and BCL-2 while negative for chromogranin, and synaptophysin which supports the synovial sarcoma diagnosis. The chromosomal translocation t(X;18) (p11.2;q11.2): SYT-SSX1 fusion can distinguish synovial sarcoma from other soft tissue tumors of soft tissue (9).

Lobectomy or pneumonectomy is the initial and most accepted method of management (8). Adjuvant or neoadjuvant chemotherapy may benefit those patients (9). Neoadjuvant chemotherapy administration is equivocal, according to recent updates, as there is no proof that the overall survival rate has improved (12–14). In high-grade lesions (G2-3), deep and >5 cm masses, and in situations where R0 resections were not possible, adjuvant radiotherapy may provide acceptable local control of residual malignant cells post-surgical resection (15,16).

The prognosis of PPSS is usually poor and depends on the stage of tumor progression with a mean survival of 5 years (2,9). Several factors determine the poor prognosis of PPSS including tumor size >5cm, increase mitotic activity (>9 mitoses per 10 High Power Field), neurovascular invasion, male sex, young adult >20 years old, and presence of SYT–SSX (2,9).

5. CONCLUSION

PPSS is a rare entity of lung malignancies with aggressive behavior and poor outcome. The presentation of those patients is usually similar to other lung malignancies, and hence biopsy or final pathological assessment is the definitive diagnosis. There is no management guideline has been proposed in the literature, and most management plans are based on surgical resection if feasible with a combination of both chemo- and radiation therapy. Our patient underwent lobectomy with adjuvant radiotherapy, and due to his comorbidities, recurrence, and metastasis, he passed away after 26 months of the initial diagnosis.

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