CASE REPORT

An Uncommon Case of Primary Synovial Sarcoma of the Lung with a Rare Intraoperative Finding

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Summary

We report a case of a 32-year-old male who was evaluated for right chest pain and cough with streaky hemoptysis. Chest X-ray and contrast-enhanced computerized tomography scan of the chest showed a large (20x16×16 cm), heterogeneously enhancing mass arising from the right lower lobe. Ultrasound-guided Tru-cut biopsy revealed spindle cell sarcoma. In immunohistochemistry, tumor cells expressed epithelial membrane antigen, CD99, and bcl-2. The final diagnosis of primary pulmonary synovial sarcoma was confirmed after positron emission tomography-computed tomography revealed that the lesion was confined to the right lung. The patient was managed by multimodal treatment. The patient underwent right anterolateral thoracotomy and lower lobectomy with staged systematic mediastinal lymphadenectomy.

Intra-operatively, he was given 2 units of packed red blood cells (RBCs). The patient developed dark-colored urine near the end of the transfusion of the second unit of packed RBCs, but without any hemodynamic instability. All the tests for possible blood transfusion reactions were negative; thus, we concluded that this event was probably secondary to tumor lysis due to the handling of the large primary lesions during surgery.

Keywords: Primary synovial sarcoma; lung sarcoma; rare lung tumor; tumor lysis urine.

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Introduction

Soft tissue sarcomas account for less than 1% of the overall human burden of malignant tumors; however, they are life-threatening. The relatively small number of cases and the great diversity in anatomic sites, biologic behaviors, and histopathologic features have made the comprehensive understanding of these disease entities difficult. Primary malignant mesenchymal tumors or soft tissue sarcomas originating from lungs are very rare. Synovial sarcomas account for 10% of total soft tissue tumors, which are more common in the upper and lower limb regions, but less common in the head and neck regions. Pulmonary sarcomas are very uncommon and comprise only 0.5% of all primary lung malignancies. Primary synovial sarcoma arising from the lung is extremely rare, with very few cases being reported (1, 2). We herein report a case of primary synovial sarcoma

of the lung arising from the right lung lower lobe that was successfully managed by a multidisciplinary team (MDT), and the patient was doing well for 24 months after completion of treatment.

Case presentation

A 32-year-old male, nonsmoker, complained of a cough with blood-stained sputum associated with right-sided chest pain for 3 days. There were no constitutional symptoms. Clinically, there was reduced air entry on the right middle and lower zones; otherwise, the rest of the examination was unremarkable. All blood investigations were done, and the values were within the normal limits. Chest X-ray, posteroanterior view, revealed a mass occupying the middle and lower zones of the right chest (Figure 1A), and contrast-enhanced computed tomography (CECT) of the chest (Figure 1B) showed a large (20×16×16 cm), heterogeneously enhancing mass with necrotic areas arising from the right lung lower lobe, with compressive atelectasis of the middle and upper lobed and pressure effect on the mediastinum and displacement of the diaphragm.

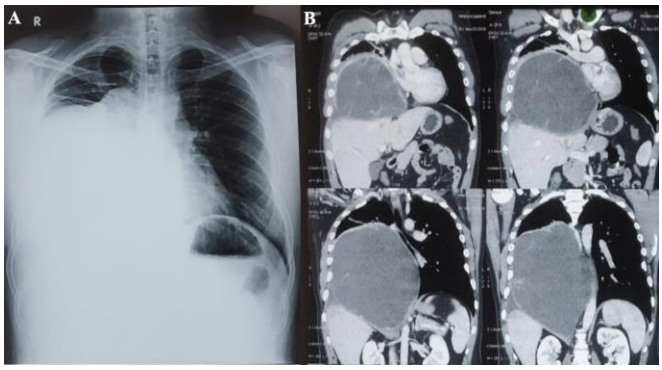

Figure 1. (A) Chest X-ray (postero-anterior view) mass occupying the middle and lower zones of the right chest. (B) Contrast-enhanced computed tomography of the chest coronal reconstructed image, showing a large, heterogeneously enhancing mass with compression and pressure effect on the mediastinum, right lung, and diaphragm.

Ultrasound-guided Tru-cut biopsy revealed pleomorphic spindle cells with hyperchromatic plump nuclei, scanty cytoplasm, mitosis, 1-2/HPF, and necrosis suggestive of spindle cell malignant tumor. The result of further evaluation with immunohistochemistry (IHC) was suggestive of synovial sarcoma (bcl-2-positive, CD99-positive, EMA-weak positive, CK7-negative, Desmin-negative, S-100-negative, SMA-negative, CD34-negative). The diagnosis of primary synovial sarcoma arising from the right lung was made based on the primary lesion confined to the right lung lower lobe revealed by fluorodeoxyglucose–positron emission tomography (PET). The case was discussed in the tumor board meeting, and the patient received two cycles of neoadjuvant chemotherapy (ifosfamide+doxorubicin-based). Because of increasing symptoms after two cycles, the patient was re-evaluated by PET-CT, showed no significant interval regression in tumor size. He underwent right postero-lateral thoracotomy followed by excision of the mass with right lower lobectomy and systematic staged mediastinal lymphadenectomy. The right upper and middle lobes were not involved and were thus preserved. Two units of packed red blood cells (RBCs) were transfused intra-operatively, but at the end of the transfusion of the second unit, the patient developed dark-colored urine, mimicking hematuria (Figure 2A), without any hemodynamic instability. The transfusion was stopped, and all the tests for possible blood transfusion reactions were negative. A peripheral blood smear was normocytic normochromic, and the urine showed only 1-2 RBCs/HPF.

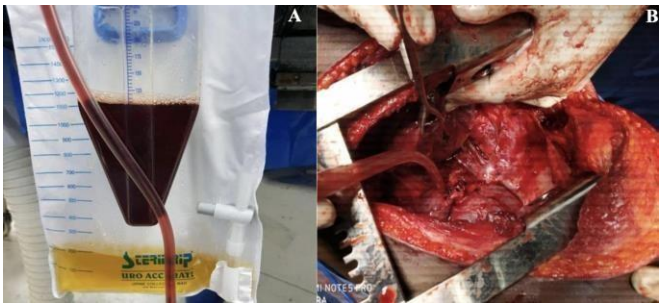

Figure 2. (A) Clear urine in the bag after catheterization and subsequent dark-colored urine (mimicking hematuria) in the collection chamber intra-operatively after blood transfusion. (B) Tumor was densely adherent to the right lung lower lobe.

It was resolved completely by the second postoperative day. Thus, it was concluded that the colored urine was probably caused by tumor lysis due to the mobilization and handling of large size tumor and preoperative chemotherapy. In our case, classical features of tumor lysis syndrome (hyperuricemia, hyperkalemia, hyperphosphatemia, and hypocalcemia) were not

present. The serum electrolytes and other blood reports were normal on the first postoperative day, and the patient remained hemodynamically stable throughout. Low-power (×10 magnification) histopathology examination showed spindle cells arranged in fascicles with infiltrative borders (Figure 3A), and high-power (×40 magnification) histopathology examination showed highly cellular spindle cells arranged in fascicles having oval to spindled nuclei and scant cytoplasm (Figure 3B), which are characteristics of synovial sarcoma. The mediastinal pleural-based nodule was involved by the tumor, but there was no lymph node involvement.

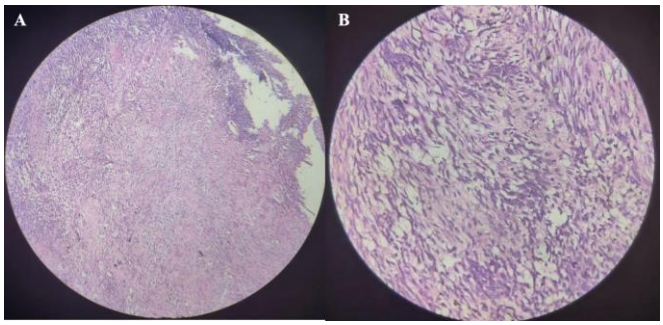

Figure 3. Histopathology images. (A) Low power (10× magnification), showing spindle cells arranged in fascicles with infiltrative borders. (B) High power (40× magnification), showing highly cellular spindle cells arranged in fascicles having oval to spindled nuclei and scant cytoplasm.

Because of the large size of the tumor, mediastinal pleural-based nodule involvement by the tumor, the MDT decided to give the patient adjuvant radiotherapy. At present, the patient has completed four cycles of adjuvant chemotherapy (ifosfamide+doxorubicin-based) and adjuvant radiotherapy (remains asymptomatic), and the latest PET-CT scan 24 months after surgery was normal.

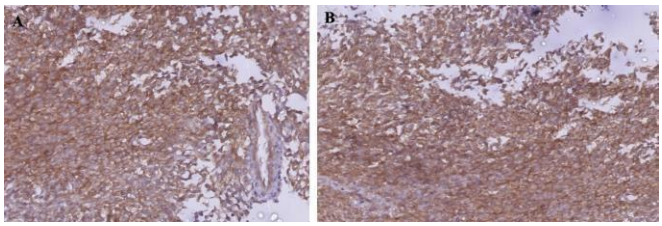

Figure 4. Immunohistochemistry images. (A) CD99-positive image. (B) bcl-2-positive image

Discussion

Although named synovial sarcoma due to its resemblance to synovium on light microscopy, it arises from mesenchymal tissue. The diagnosis is established only after sarcoma-like primary lung malignancy and metastatic sarcoma have been excluded. Metastasis from extra-pulmonary sarcoma is more common than primary pulmonary sarcomas (1, 2). Synovial sarcomas are common in young adults between 25 and 35 years and have a male predominance. They are highly aggressive malignant neoplasms, and they are not related to smoking or asbestos exposure. Chest-ray, CECT, PET scan, and IHC are helpful tests for diagnosing synovial sarcomas. All these investigations were done on our patient to confirm the diagnosis. However chromosomal analysis was not performed on our patient because of the lack of this facility in our hospital.

Cytogenetically, nearly all synovial sarcomas contain characteristic chromosomal translocation, t(X; 18). Histologically, there are four subtypes of synovial sarcomas: monophasic spindle cell, monophasic epithelial cell, biphasic, and poorly differentiated (3, 4). Our case was characterized by the presence of spindle cell sarcoma, and tumor cells were negative for cytokeratin 7, Desmin, S-100, SMA, and CD34 and positive for epithelial membrane antigen, CD99, and bcl-2 (Figure 4). Thus, the diagnosis of primary synovial sarcoma of the right lung was confirmed. In our patient, two units of packed cells were transfused intra-operatively, and at the end of the transfusion of the second unit, dark-colored urine was observed, but the patient had no hemodynamic instability. Transfusion was stopped immediately. Intra-operative hematuria is a critical event; thus, we evaluated thoroughly and double-checked all blood cross-matching results; peripheral smear showed no hemolysis, and urine microscopy and cytology for malignant cells were negative. Post-operatively, we reviewed the patient's CT scans, but there was no urinary pathology such as urinary calculi or malignancy. The urine became completely clear by the end of the second postoperative day. A literature search revealed that very few reports showed that intraoperative hematuria after transfusion of cell saver blood in orthopedic surgery disappeared after 7 hours of surgery (5). Huh, et al. (6) reported a similar case. In our

case, we have not transfused cell-saver blood. The prognosis of patients with primary pulmonary sarcoma is poor (overall 5-year survival rate, 50%). Male sex, age >20 years, tumor size >5 cm, high grade, a high number of mitotic figures (>10/10 HPFs), neurovascular invasion, and SYT-SSX1 variants are associated with poor prognosis (7). In our case, the patient is male, >20 years, and with a tumor size >5 cm. The main prognostic factor is to achieve a complete resection, which was done in our case and confirmed by histopathology examination (Figure 3A and B). There is no standardized therapy because the rarity of such tumors has not permitted any controlled trials of adjuvant chemotherapy. The majority are treated with surgery alone or surgery with adjuvant chemotherapy and adjuvant radiotherapy. Synovial sarcomas are chemo-sensitive to ifosfamide and doxorubicin, with an overall response rate of approximately 24% (8, 9). The primary synovial sarcoma of the lung is an extremely rare entity. Intra-operative dark-colored urine is an equally rare finding that may be attributed to tumor mobilization during surgery, and as such, it should not be a cause of alarm if the patient remains hemodynamically stable. The MDT approach has a vital role in patient management.

Conclusion

Primary synovial sarcoma of the lung is a very rare tumor that requires an MDT approach, and multiple modality treatment with complete excision is the mainstay treatment.

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Compliance with Ethical Standards: This study was conducted in compliance with ethical standards.

Informed consent: Written informed consent for the publication of clinical details and clinical images was obtained from the patient.

Author contributions

Conceptualization (Equal); Data curation (Eqaul); Formal analysis: JAA(Lead); Funding acquisition (Equal); Investigation (Equal); Methodology (Equal); Project administration: RR(Lead); Resources: (Equal); Software (Equal); Supervision: RR (Lead); Validation (Equal); Visualization (Equal); Writing – original draft: RCM(Lead);Writing – review & editing: RR(Lead).

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