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Primary Synovial Sarcoma of Lung

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Synovial sarcoma (SS) is a highly malignant tumor that accounts for 10% of all soft-tissue sarcomas. Primary SS arising from the lung is extremely rare, and the prognosis is poor. We report a case of pulmonary SS presenting with a mass lesion invading the right upper and middle lobes, extending to the mediastinum and the chest wall. After tru-cut biopsy, surgical resection was performed. The final diagnosis was SS (biphasic type) based on histological and immunohistochemical findings. There are no guidelines for optimal treatment due to the rarity of these tumors. Current treatment includes surgery and adjuvant chemotherapy and/or radiotherapy.

Key words: 1. Synovial sarcoma
2. Lung
3. Immunohistochemistry
4. Adjuvant therapy

CASE REPORT

Synovial sarcoma (SS) is a highly malignant tumor that occurs mainly in adolescents and young adults and is usually seen in the extremities [1]. However, primary synovial sarcoma arising from the lung is extremely rare, accounting for 0.3% to 1.3% [2]. It is closely associated with smoking [3]. Besides clinical evaluation and imaging methods for definitive diagnosis, immunohistochemical examination is needed. We report a case of pulmonary SS with a history of heavy smoking.

A 69-year-old man was admitted to the hospital with right-sided chest pain. His physical examination revealed no pathology. No peripheral lymphadenopathy was detected. The patient was a heavy smoker with a history of smoking 120 packs/yr. A right middle zone opacity was detected on the chest X-ray. Computed tomography (CT) of the chest revealed a 60×56×70 mm mass lesion invading the right upper and middle lobes and extending to the mediastinum and the chest wall (Fig. 1). Tru-cut biopsy was taken from the lesion, and histological examination was suspicious about malignancy. Positron emission tomography/CT detected an increased F18-fluorodeoxyglucose uptake (maximum standardized uptake value, 8.2) only in the mass.

Surgical exploration was decided upon, and right bilobectomy superior bronchoplasty with mediastinal lymph node dissection was performed. The lobectomy material was 20×11×5.5 cm in diameter, and macroscopic examination revealed an 11-cm, well-demarcated, brownish, necrotic mass in continuity with the bronchial tree and adjacent to the pleura (Fig. 2). Histopathological examination showed long bundles of spindle cells with no pleomorphism and gland-like structures including single-row epithelium, to be more prevalent in the tumor periphery (Fig. 3). The dissected lymph nodes and

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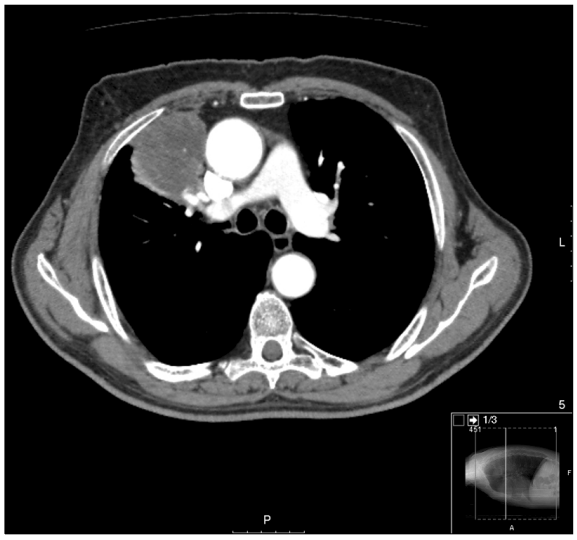

Fig. 1. Computed tomography of the chest showing a mass lesion invading the right upper and middle lobes and extending to the mediastinum and the chest wall.

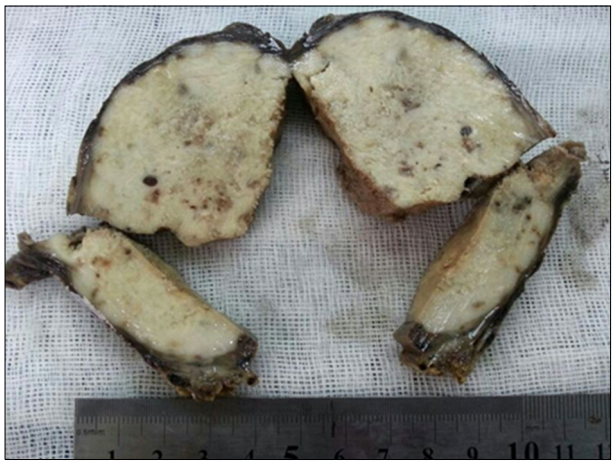

Fig. 2. Macroscopic examination showing an 11-cm, well-demarcated, brownish, necrotic mass in continuity with the bronchial tree and adjacent to the pleura.

pleura were negative for tumor cells. By immunohistochemistry, the spindle cells were positive for B-cell lymphoma 2 (Bcl-2) in the scattered foci and negative for CD99, Wilm's tumor-1, actin, caldesmon, CD34, S-100, pan-cytokeratin, and endomysial antibodies (EMA) (Fig. 4). The epithelium of gland-like structures was positive for EMA and pan-cytokeratin (Fig. 4). The epithelial groups at the periphery of the tumor were positive for thyroid transcription factor

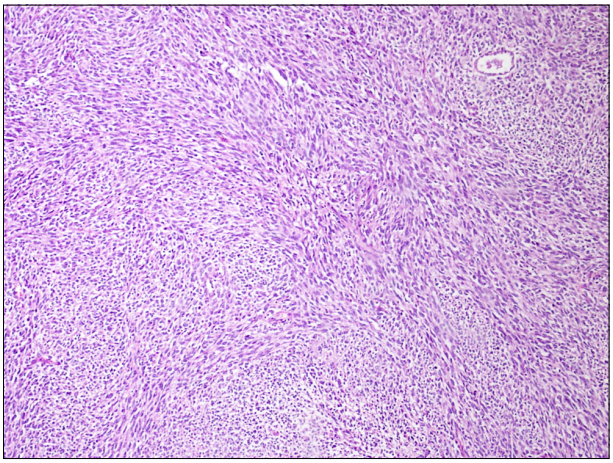

Fig. 3. Long bundles of spindle cells and the epithelial component forming gland-like structures (H&E, ×200).

1. As a result of morphological and immunohistochemical findings, spindle cell SS (biphasic type) was diagnosed. Following surgery, we planned 4 cycles of adjuvant chemotherapy followed by radiotherapy. He received 4 cycles of adjuvant chemotherapy and then completed the courses of radiotherapy on June 2013. He is still in remission at the 1-year follow-up after surgery.

DISCUSSION

Synovial sarcoma is a rare mesenchymal tumor, first described by Simon in 1865. It accounts for 10% of all soft-tissue sarcomas and is slightly more common in men [4]. Four histologic subtypes are described: biphasic, monophasic (spindle), monophasic epithelial, and poorly differentiated (round cell) tumor [5]. Monophasic tumor is the most commonly observed subtype. The biphasic subtype is easily diagnosed on the basis of the presence of both epithelial and spindle cells. Monophasic subtype can be mixed up with other types of sarcoma, and therefore, immunohistochemistry is essential for differential diagnosis. Our case was characterized by sarcoma composed of spindle cells and epithelial cells on histopathological examination; epithelial cells were positive for EMA and pan-cytokeratin, and spindle cells were positive for Bcl-2.

Although the cytomorphologic and immunohistochemical features of SS are sufficiently characteristic for diagnosis,

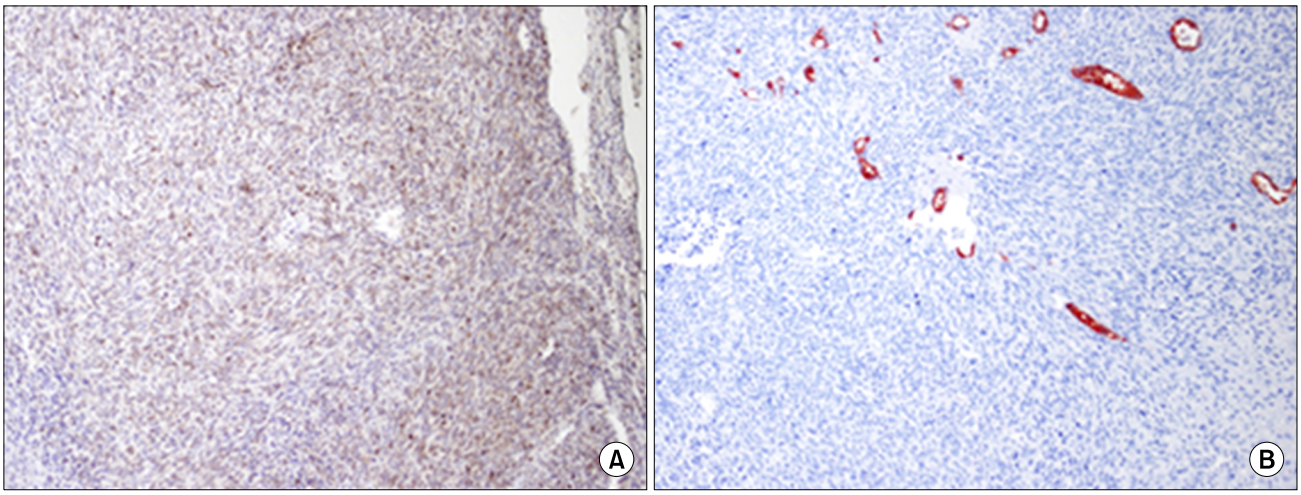

Fig. 4. (A) Photomicrograph of spindle cells showing scattered focal positive nuclear staining with B-cell lymphoma 2 (Bcl-2, ×200). (B) Photomicrograph showing endomysial antibodies (EMA)-positive cytoplasmic staining of the epithelial component (EMA, ×200).

clinical correlation is also essential for an accurate diagnosis of primary SS of the lung. In our case, the diagnosis of the primary SS of the lung was also based on the absence of the tumor at any other site of the body.

Cytogenetics plays an important role in the diagnosis of SS. Both monophasic and biphasic forms are characterized by a reciprocal chromosomal translocation (x; 18) (p11.2; q11.2) which results from the fusion of the SYT gene on chromosome 18 to either of two genes, SSX1 and SSX2 in the region xp11 [6]. Despite its high sensitivity, molecular testing is not essential for the diagnosis of SS. In our case, molecular testing was not performed due to the certain diagnosis of SS on the basis of clinical, histological, and immunohistochemical findings.

Sixty six percent of primary pulmonary SS are centrally located and present with obstructive pneumonia, hemoptysis, dyspnea, cough, and fever [7]. Peripheral tumors are rare. SS can also arise from the pleura. However, primary SS of the pleura is also very rare and there are a limited number of reported cases [8]. Our case presented with a centrally located tumor and a complaint of chest pain. The tumor was adjacent to the pleura, but there was no pleural involvement microscopically.

Surgical resection with tumor negative margins is the primary treatment. Prior to surgery, the primary focus of SS or metastasis should be excluded by imaging procedures. These tumors are highly aggressive, and the overall prognosis is poor [9]. There are no guidelines for optimal treatment due to the rarity of these tumors. Current treatment includes adjuvant chemotherapy and/or radiotherapy [6,7]. Our patient underwent right bilobectomy and mediastinal lymph node dissection and then, received four cycles of adjuvant chemotherapy and radiotherapy postoperatively.

In conclusion, primary pulmonary SS is an extremely rare and aggressive tumor. Immunohistochemical staining and clinical imaging is necessary for definitive diagnosis. Surgical resection along with adjuvant chemo-radiotherapy is the currently accepted therapy.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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