

Primary pulmonary synovial sarcoma: a rare neoplasm

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Abstract

Primary pulmonary synovial sarcoma is an extremely rare tumor with an unknown cause. The diagnosis is established after other primary lung malignancies or metastatic extrathoracic sarcoma have been excluded. We report the case of a 69-year-old man who presented with a well-defined mass in the right upper lobe on a chest X-ray. A video-assisted thoracoscopic surgery (VATS) right upper lobectomy was performed. Immunohistochemically, neoplastic cells were positive for vimentin, CD56 and Bcl-2, and focally positive for CD99, epithelial membrane antigen and cytokeratin 7 and 19. The cytogenetic study revealed a SYT genetic reassortment. So, the final pathological diagnosis was primary pulmonary synovial sarcoma.

Key words: synovial sarcoma, lung mass, immunohistochemistry.

Streszczenie

Pierwotny płucny mięsak maziówkowy to niezwykle rzadko spotykany nowotwór o nieznannej etiologii. Rozpoznanie ustala się po wykluczeniu innych pierwotnych nowotworów złośliwych płuc oraz przerzutów mięsaka spoza klatki piersiowej. Przedstawiamy przypadek 69-letniego pacjenta, u którego badanie rentgenowskie uwidocznilo obecność wyraźnie odgraniczonej masy w górnym płacie prawego płuca. Przeprowadzono lobektomię górną prawą z zastosowaniem techniki wideotoraskopowej (VATS). Wyniki badania immunohistochemicznego komórek nowotworowych były dodatnie dla wimentyny, CD56 i Bcl-2, a ogniskowo dodatnie dla CD99, antygenu błony nabłonkowej oraz cytokeratyn 7 i 19. Badanie cytogenetyczne ujawnilo reasortację genetyczną SYT. W związku z powyższym patologię zdiagnozowano ostatecznie jako pierwotny płucny mięsak maziówkowy.

Słowa kluczowe: mięsak maziówkowy, guz płuca, immunohistochemia.

Introduction

Primary pulmonary synovial sarcomas are extremely rare neoplasms with an unknown cause originating from mesenchymal tissue and accounting for 10% of soft tissue sarcomas [1].

Although most synovial sarcoma tumors are located in soft tissue, especially near large joints of the extremities, they also can occur in numerous situations unrelated to joint structures. Thoracic involvement of the synovial sarcoma is rare, and only a few cases have been reported in the literature.

Case report

We report the case of a 69-year-old asymptomatic man, a heavy smoker with no clinically relevant family or personal history.

The patient was admitted to our institution for study of a solitary pulmonary nodule found after routine chest X-ray, not present in previous examinations. Computed tomography (CT) scan showed a 23 mm multilobulated nodule in the right upper lobe with peripheral calcification (Fig. 1 A).

No significant mediastinal lymph nodes were observed. The standardized uptake value of the pulmonary nodule in positron emission tomography-computed tomography (PET-CT) scan was 1.5 g/ml. Bronchoscopy did not reveal any endobronchial lesion. Transbronchial fine-needle aspiration biopsy was performed. Nevertheless, a diagnosis was not established by this technique.

In order to resect the nodule, a video-assisted thoracoscopic surgery (VATS) right upper lobectomy with systematic lymph node dissection was performed. Pathological examination revealed a well-defined tumor, not encapsulated, with spindle cells (Fig. 1 B). Immunohistochemically, neoplastic cells were positive for vimentin, CD56 and Bcl-2, and focally positive for CD99 (Fig. 1 C), epithelial membrane antigen and cytokeratin 7 and 19. Cytogenetic study by reverse transcriptase-polymerase chain reaction revealed a SYT genetic reassortment. The final pathological diagnosis was primary pulmonary synovial sarcoma. No spread to nearby lymph nodes was noted. Postoperative evolution was without complications and the patient was discharged 7 days after surgery.

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Discussion

Synovial sarcoma is a rare pulmonary tumor accounting for 10% of soft tissue tumors. However, a few cases of this kind of neoplasm have been described [2]. It typically occurs in adolescents and young adults. More than 90% of synovial sarcomas are located in extremities. Pulmonary synovial sarcoma constitutes between 0.1% and 0.5% of all lung neoplasms.

Histologically, primary pulmonary synovial sarcoma can be classified into four categories: biphasic, monophasic fibrous (spindle cells), monophasic epithelial and poorly differentiated types. Macroscopically, these tumors are well circumscribed and not encapsulated, with a very variable size ranging from 0.6 to 27 cm (mean: 6.8). They may present aggressive behavior, infiltrating nearly all structures. Immunohistochemistry has an important role in the diagnosis. Synovial sarcomas are positive for cytokeratin 7 and 19, EMA, Bcl-2, CD99 and vimentin. They are usually negative for S-100, CD-34, desmin, actin and vascular tumor markers [3–5]. Recent cytogenetic studies have taken a leading role for definitive diagnosis of synovial sarcoma, identifying a translocation $t(X;18)(p11.2;q11.2)$ resulting from fusion of the SYT gene on chromosome 18 to SSX1 or SSX2 on chromosome X [2].

Differential diagnosis includes other malignant extrathoracic tumors such as fibrosarcomas, carcinosarcomas, leiomyosarcomas or hemangiopericytomas. Metastatic disease must be ruled out, especially the monophasic type.

There is no standardized therapy for patients with primary pulmonary synovial sarcoma. However, surgical complete resection remains the main strategy in these patients. In advanced forms or unresectable tumors doxorubicin and ifosfamide based chemotherapy can be used [6].

The prognosis for patients with primary pulmonary synovial sarcoma is poor, with an overall 5-year survival rate of 50%. Negative prognostic factors are tumor size, male gender, extensive tumor necrosis, higher histological grade, mitotic rate and neurovascular invasion. The expression of SYT-SSX1 variants has been associated with worse behavior.

Disclosure

Authors report no conflict of interest.

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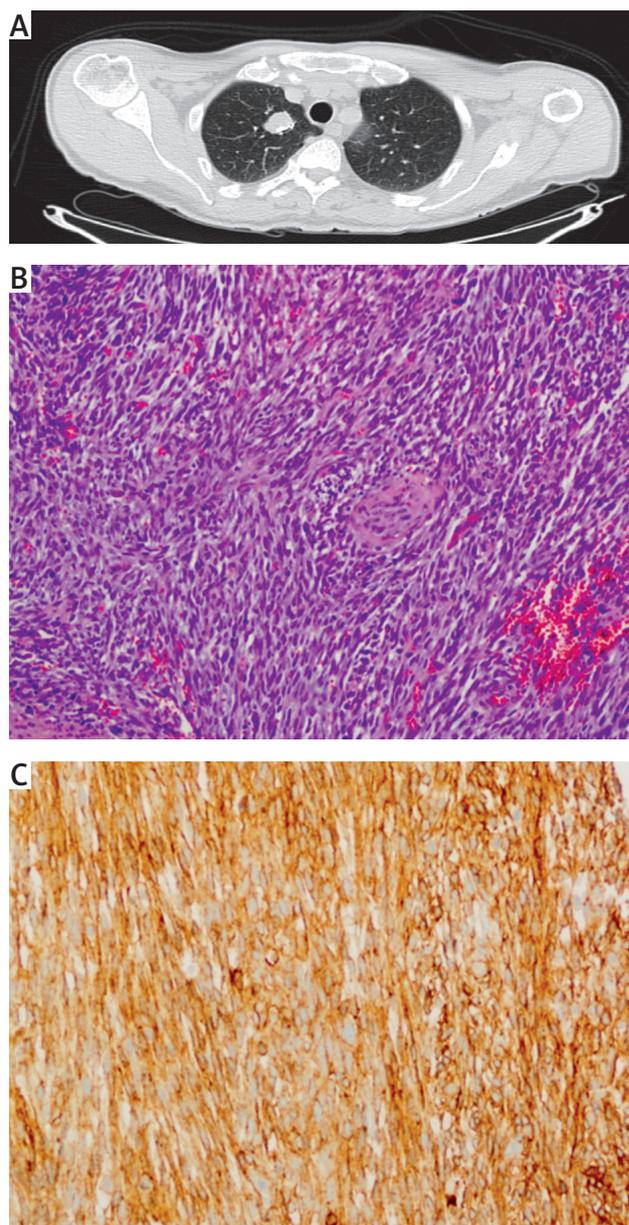


Fig. 1. A) Chest CT scan shows a multilobulated nodule in the right upper lobe with peripheral calcification. B) Spindle cell proliferation organized in irregular fascicles, without epithelial component in tumor cells (H/E 40 \times). C) Intense immunohistochemical expression of CD99 membrane marker in tumor cells (40 \times)