Metastatic Monophasic Synovial Sarcoma of the Pleura

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Pleural metastasis of synovial sarcoma that originally developed in the soft tissue is a very rare entity. We report here the detailed clinical features of such a case. An asymptomatic 25-year-old female, with a history of a resected synovial sarcoma in her left brachial muscle and pulmonary metastasectomy of the right lung, presented a small nodule in the periphery of the left lung on a routine chest-computed tomography. Exploratory thoracoscopy was then performed. A soft flat red tumor approximately 2 cm in diameter was shown on the pleura of the lingula, mimicking a blood clot on the pleura. The tumor was removed by partial resection of the lung. The mass lay in the pleura and did not seem to invade the lung parenchyma macroscopically. Intraoperative frozen sectioning evidenced metastatic synovial sarcoma. Many small patchy red lesions were also found on the visceral pleura of the lung and parietal pleura of the diaphragm. We diagnosed unresectable pleural metastases of synovial sarcoma and finished the operation after sampling another pulmonary pleural lesion. The patient then underwent ifomide-based chemotherapy and survived for 3 years after her initial surgery. Postoperative histopathological examination revealed a solid and bundle-like proliferation of a short spindle cell tumor with a monophasic pattern, which was diagnosed as a metastatic pleural synovial sarcoma. A hemangiopericytomatous pattern was also seen; therefore the lesion looked like a blood clot because of its rich vascularity. (Ann Thorac Cardiovasc Surg 2007; 13: 258–261)

Key words: pleura, neoplasm, synovial sarcoma, surgery, metastasis

Introduction

The lung is the most common site of metastasis in patients with soft tissue sarcoma, even those with synovial sarcoma. However, the pleural metastasis of synovial sarcoma that originally developed in the extremities is a very rare entity; indeed, to our knowledge it has not been previously reported. We report here detailed clinical features of such a case with reference to some of the relevant literature.

Case

An asymptomatic 25-year-old female, with a history of a resected synovial sarcoma in her left brachial muscle 3 months previously, presented a right lung mass 1 cm in diameter detected on follow-up computed tomography (CT). She had undergone chemotherapy with ifomide alone, ifomide and adriamicin, and cisplatin and adriamicin, in that order, preoperatively, and with ifomide postoperatively.

Three months later, a reevaluation on chest CT demonstrated the same lung mass. The size had not increased. Pulmonary metastasectomy of the right lung was performed 6 months after the main tumor resection. The tumor was a nodular lesion in the lung that was palpated as elastic hard and was approximately 1 cm in diameter. Histologically, the lung mass evidenced metastatic synovial sarcoma. Four months after the metastasectomy, a small lung mass was again detected in the periphery of the lingula on follow-up CT (Fig. 1A). Exploratory thoracoscopy was then performed.

A soft flat red tumor approximately 2 cm in diameter was shown on the pleura of the lingula, mimicking a blood clot on the pleura. The tumor was removed by partial resection of the lung. The mass lay in the pleura and did not seem to invade the lung parenchyma macroscopically. Intraoperative frozen sectioning evidenced metastatic synovial sarcoma. Many small patchy red lesions were also found on the visceral pleura of the lung and parietal pleura of the diaphragm. We diagnosed unresectable pleural metastases of synovial sarcoma and finished the operation after sampling another pulmonary pleural lesion. The patient then underwent ifomide-based chemotherapy and survived for 3 years after her initial surgery. Postoperative histopathological examination revealed a solid and bundle-like proliferation of a short spindle cell tumor with a monophasic pattern, which was diagnosed as a metastatic pleural synovial sarcoma. A hemangiopericytomatous pattern was also seen; therefore the lesion looked like a blood clot because of its rich vascularity. (Ann Thorac Cardiovasc Surg 2007; 13: 258–261)

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Clot on the pleura (Fig. 2A). The tumor was removed by a partial resection of the lung. The mass lay in the pleura and seemed not to invade the lung parenchyma macroscopically. Intraoperative frozen sectioning evidenced metastatic synovial sarcoma. Many small patchy red lesions were also found on the visceral pleura of the lung and the parietal pleura of the diaphragm (Fig. 2B). We diagnosed unresectable pleural metastases of synovial sarcoma and finished the operation after sampling another pulmonary pleural lesion. A postoperative examination of the specimen revealed a flat polypoid tumor with a wide pedicle (Fig. 3A). Although the tumor was partially invading into the lung parenchyma, the lesion was mainly lodged in the pleura.

A histopathological examination revealed a solid and bundle-like proliferation of a short spindle cell tumor with a monophasic pattern, and it was diagnosed as a metastatic pleural synovial sarcoma (Fig. 3B). A hemangiopericytomatous pattern was also seen; therefore, the lesion looked like a blood clot because of its rich vascularity. One month later, multiple pleural lesions of the lingula were growing in follow-up CT (Fig. 1B). However, after the patient underwent repeated ifomide-based chemotherapy, these lesions had disappeared completely, and she survived without recurrence for 42 months after her initial surgery.

**Discussion**

Synovial sarcoma is an uncommon malignant soft tissue neoplasm that mostly occurs in the extremities, especially in periarticular regions. Synovial sarcoma can arise from...
the organs without a synovial membrane, such as the pleura. The term "synovial" sarcoma was given because of the synovial differentiation of the tumor that is believed to originate from multipotential mesenchymal cells. Synovial sarcoma is predilected in those aged from adolescence to their 40s. Detection of the synovial sarcoma-specific transcriptions (SYT-SSX1 or SYT-SSX2) by polymerase chain reaction is known to be helpful for diagnosis.

Metastatic synovial sarcoma of the pleura is an extremely rare entity and to our knowledge has not previously been reported. Its appearance is peculiar, mimicking a blood clot just fallen from a thoracoscopy wound onto the lung. Small lesions also look like patchy hemorrhages of the lung surface.

Pleural primary synovial sarcomas, which originally developed in the pleura, have been reported. Pleural synovial sarcoma tends to form a large mass in the pleural cavity, and diagnosis is sometimes very difficult because synovial sarcoma is histologically similar to sarcomatous mesothelioma. In our case, its diagnosis was histologically confirmed from a specimen of the left arm and the right lung. Small lesions also look like patchy hemorrhages of the lung surface.

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Pulmonary metastasectomy of the soft tissue sarcoma is efficient to elongate survival in selected patients with small-sized, limited resectable numbers of pulmonary nodules, and with a longer disease-free interval or low-grade histological type of the primary disease. Pleural metastasis should be treated according to the same criteria; however, in our case numerous small pleural lesions were found; only the largest main lesion was resected, and the patient was then treated postoperatively with chemotherapy.

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References