**Malignant Solitary Fibrous Tumor Arising in the Right Buttock Associated with Metastatic Parietal Pleural and Intrapulmonary Tumors in Addition to Pleural Effusion**

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A malignant solitary fibrous tumor arising in the right buttock associated with metastatic parietal pleural and intrapulmonary tumors and pleural effusion was found in a 59-year-old man. A chest computed tomogram revealed three tumors attached to the parietal pleura with rib destruction, and a tumor in the left lower lung field. Histologically, the tumors of the buttock and parietal pleura were characterized by proliferation of bundles of spindle-shaped or oval cells separated by wavy hyalinized collagen tissue with no expression of cytokeratin, S-100 protein, muscle actin or epithelial membrane antigen, but these cells weakly expressed CD34 and strongly expressed vimentin.

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**Key words:** metastatic tumor, mitotic activity, high cellularity, CD34, nuclear pleomorphism, vimentin

**Introduction**

Solitary fibrous tumors are neoplasms that usually involve the pleura and less frequently occur in a variety of other sites including the peritoneum (1, 2), liver (3), upper respiratory tract (4, 5) mediastinum (6) and orbit (7). It is very rare for this neoplasm to involve the buttocks (8) or the extremities (9). It has been reported that 12–23% of patients with solitary fibrous tumor have recurrence and metastasis (10–12). This report describes a 59-year-old man with a malignant solitary fibrous tumor arising in the right buttock associated with metastatic parietal pleural and intrapulmonary tumors as well as pleural effusion.

**Case Report**

A 59-year-old non-smoking man was admitted for evaluation of a tumor shadow in the left middle lung field and pleural effusion in June 1996 (Fig. 1). He had been in good health and had a large mass (10×8 cm) on his right buttock for 40 years (Fig. 2). On admission, arterial blood gas analysis revealed slight hypoxemia (pH 7.37, partial pressure of oxygen (PaO₂) 75.0 Torr, partial pressure of carbon dioxide (PaCO₂) 36.5 Torr, HCO₃ 21.1 mmol/l). The results of blood counts and blood chemistry tests were almost normal and tumor markers including carcinoembryonic antigen (CEA), cytokeratin 19 fragment (CYFRA 21-1) and neuron specific enolase (NSE) were within normal levels.

A computed tomogram (CT) of the chest revealed three tumors attached to the parietal pleura with rib destruction, a tumor of the left lower lung field and left pleural effusion (Fig. 3). A ⁹⁹ᵐTc-MDP scintigram showed three abnormal accumulations on the ribs corresponding to the tumors attached to the parietal pleura (Fig. 4). Analysis of the pleural effusion indicated an exudative inflammation with increased lymphocyte levels but normal CEA levels.

Because attempts of both transbronchial lung biopsy and transcutaneous needle aspiration cytology were of no diagnostic value, a posterolateral thoracotomy was performed, which revealed multiple disseminated small tumors on both visceral...
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Figure 1. A chest roentgenogram on admission showed a tumor shadow in the left middle lung field and pleural effusion.

and parietal pleuras and bloody effusion in addition to three tumors attached to the parietal pleura. We thus performed resection of one of the those tumors with a partial resection of thoracic wall. At the same time, we resected the intrapulmonary tumor and the mass of his right buttock.

Histologically, sections from the buttock mass showed proliferation of interlacing bundles of spindle-shaped or oval cells separated by thick strands of rope-like collagen and were diagnosed as solitary fibrous tumor (Fig. 5A). However some parts of the buttock mass had high cellularity, nuclear pleomorphism and mitotic activity (more than 4 mitoses/10 high-power fields), but no hemorrhage or necrosis (Fig. 5B). Histological findings of the parietal pleura and lung tumor were similar to those of the buttock mass with a high cellularity, nuclear pleomorphism and mitotic activity (Fig. 5C, D). In addition, alveolar pneumocytes and small bronchioles became entrapped within tumor cell proliferation in the intrapulmonary tumor (Fig. 5D). Immunohistochemical studies showed staining of tumor cells by vimentin and negative stain with cytokeratin, S-100 protein, muscle actin and epithelial membrane antigen (EMA) (Table 1). In addition, these cells weakly expressed CD34 (Fig. 6). The features were considered to be typical of the solitary fibrous tumor arising in the buttock and metastatic to the lung and pleura. The patient is still alive with no enlargement of the tumors or recurrence of the pleural effusion six months after diagnosis despite receiving no treatment because he refuses both chemotherapy and radiotherapy.

Figure 2. A computed tomogram showed a giant tumor (arrow) in the right buttock.

Discussion

Solitary fibrous tumor, which was first reported as being different from diffuse mesothelioma in 1931 by Klemperer and Rabin (13), usually arises in the pleura. Solitary fibrous tumor, although rare, has been found to occur at a variety of sites including the peritoneum (1, 2), liver (3), upper respiratory tract (4, 5) mediastinum (6) and orbit (7). To our knowledge, this is the second report of solitary fibrous tumor arising in the buttock. Suster et al (8) first reported a slow-growing soft tissue nodule in the buttock that had been present for 10 years before diagnosis. The present case had a similar clinical course and the buttock tumor had been present for 40 years.

Histologically, solitary fibrous tumor is characterized by a proliferation of interlacing bundles of spindle-shaped or oval cells demonstrating a variety of growth patterns, including storiform, herringbone and hemangiopericytoma-like formations. In some areas of solitary fibrous tumor, there are more advanced degrees of collagenization and spindle-shaped or oval cells separated by strands of rope-like collagen, which is referred to as the “patternless pattern” (8, 14) resembling the histological findings in the present case. However some parts of the buttock mass had a high cellularity, nuclear pleomorphism and mitotic activity and we speculate that those parts of the buttock mass metastasized to the parietal pleura and lung. Primary intrapulmonary solitary fibrous tumors have been histologically reported to consist of alveolar pneumocytes and small broncholes entrapped within tumor cell proliferation (15). The intrapulmonary tumor in our case was a metastatic lesion and the histological findings were similar to those of the primary lesion. Malignant fibrous histiocytoma (MFH) is a very important entity to be excluded in establishing the differential diagnosis of solitary fibrous tumor arising in the soft tissue. The pathological findings of MFH showed storiform growth pattern, pleomorphic malignant giant cells, spindle-shaped cells with cytologic atypia and mitotic activity, which allowed easy differentiation from the solitary fibrous tumor.

Immunohistochemical studies showed positive staining of
Figure 3. A computed tomogram of the chest revealed three tumors attached to the parietal pleura with rib destruction, a tumor in the left lower lung field and left pleural effusion.

tumor cells with vimentin and no stain with cytokeratin, S-100 protein, muscle actin and EMA, in addition to only weak expression of CD34. The CD34 monoclonal antibody is selectively expressed on human hematopoietic cells and vascular endothelium. It has been reported that the spindle-shaped or oval cells of solitary fibrous tumor are stained with CD34 (2, 7, 8, 16, 17). However CD34 positivity is not specific for solitary fibrous tumor since it is observed in dermatofibrosarcoma protuberans, neurofibromas and schwannomas, but not in sarcomatoid mesothelioma (7). In addition, not all solitary fibrous tumors have CD34 expression (2, 7, 8, 16, 17). There are many reports that the spindle-shaped or oval cells of solitary fibrous tumors do not express cytokeratins or other epithelial markers (2, 8, 15–20). Thus solitary fibrous tumors originate...
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Figure 4. A $^{99m}$Tc-MDP scintigram showed three abnormal accumulations on the ribs corresponding to the tumors attached to the parietal pleura.

Table 1. Immunohistochemical Findings of the Tumor Cells in the Buttock, Parietal Pleura and Lung

<table>
<thead>
<tr>
<th>Location of the tumor</th>
<th>Buttock</th>
<th>Parietal pleura</th>
<th>Lung</th>
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<tr>
<td>CD34</td>
<td>±</td>
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<td>±</td>
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<tr>
<td>Vimentin</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Cytokeratin</td>
<td>–</td>
<td>–</td>
<td>–</td>
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<tr>
<td>S-100</td>
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<td>–</td>
<td>–</td>
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<tr>
<td>Muscle actin</td>
<td>–</td>
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<td>EMA</td>
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from non-mesothelial cells and cytokeratins have been reported to be useful in distinguishing them from sarcomatoid mesothelioma. But since there is a recent report of a solitary fibrous tumor with expression of cytokeratin (21), combination of using cytokeratin and CD34 should be effective for the differential diagnosis between solitary fibrous tumor and sarcomatoid mesothelioma. Vimentin has been reported to be almost always positive in this disease, but this finding is not of diagnostic value, because many mesenchymal or epithelial neoplasms also have positive expressions (8, 15–18, 20, 21).

England et al (12) reviewed 223 cases of solitary fibrous tumor of the pleura and classified them as malignant using the histological criteria of increased cellularity, nuclear pleomorphism, mitosis of more than four figures per 10 high-power fields, hemorrhage and necrosis. It has been reported that 12–23% of patients with solitary fibrous tumor have recurrence or metastasis (10–12). The present case, with metastatic tumors in the parietal pleura and lung, showed increased cellularity, nuclear pleomorphism and mitoses of more than four figures per 10 high-power fields, but not hemorrhage or necrosis. Many solitary fibrous tumors of the pleura have been reported to have pleura-based pedicles (10, 12) and no abnormality of the rib adjacent to the tumor (18). Although there was a report of double primary solitary fibrous tumor of the pleura and retroperitoneum (22), we diagnosed the present case as malignant solitary fibrous tumor arising in the right buttock and metastasizing to the parietal pleura and lung because of the histological findings and three tumors with rib destruction. Solitary fibrous tumors with pleural effusion have been reported to be rare (12, 20). England et al (12) revealed that one-third of the malignant solitary fibrous tumor group had pleural effusion. We suggest that pleural effusion should be included as one of the criteria of malignant behavior of solitary fibrous tumors. It has been recently reported that quantitative DNA flow cytometry is useful for differentiation between benign and malignant solitary fibrous tumor. el-Naggar et al (23) and Domingo et al (24) demonstrated that a diploid pattern of DNA could be considered a favorable prognostic sign.

More than one-half of solitary fibrous tumor patients are asymptomatic, but the symptoms and signs associated with the solitary fibrous tumor of the pleura are chest pain, cough, dyspnea, hypoglycemia (25), finger clubbing and hypertrophic pulmonary osteoarthropathy (26). The present patient, who had multiple pulmonary lesions from a solitary fibrous tumor arising in the buttock had no symptoms or signs.

Benign solitary fibrous tumors have been treated by simple resection. However, tumors classified as malignant show frequent recurrence and thus lymph node resection should be performed. It has been reported that malignant solitary fibrous tumors can be treated by chemotherapy or radiotherapy but there is no evidence that these treatments are effective in tumor control (12). The present patient is still alive with no enlargement of the tumors or recurrence of pleural effusion six months after the diagnosis despite receiving no treatment because he refuses chemotherapy and radiotherapy. We suggest that it is not necessary for the patients with malignant solitary fibrous tumors to be treated aggressively with chemotherapy or radiotherapy because this disease is a slow-growing tumor.
Figure 5. Histological findings of the right buttock mass showed a proliferation of interlacing bundles of spindle-shaped or oval tumor cells separated by thick strands of rope-like collagen (A: HE stain, ×200). Some parts of the buttock mass had a high cellularity, nuclear pleomorphism and mitotic activity (arrow), but no hemorrhage or necrosis (B: HE stain, ×400). Histological findings of the parietal pleura and lung tumor were similar to those of the buttock mass with high cellularity, nuclear pleomorphism and mitotic activity (C, D: HE stain, ×200). In the intrapulmonary tumor, alveolar pneumocytes and small bronchioles became entrapped within tumor cell proliferation (D).

Figure 6. Immunohistochemical finding showed positive staining of tumor cells with CD 34 (arrows) as well as vascular endothelium (Immuno stain, ×200).

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References


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