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CASE REPORT

Cutaneous Metastatic Undifferentiated Pleomorphic Sarcoma from a Mediastinal Sarcoma

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Undifferentiated pleomorphic sarcoma, known as malignant fibrous histiocytoma, is a malignant neoplasm that arises in both soft tissue and bones. In 2002, the World Health Organization declassified malignant fibrous histocytoma as a formal diagnostic entity and renamed it 'undifferentiated pleomorphic sarcoma not otherwise specified.' It most commonly occurs in the lower extremities and rarely metastasizes cutaneously. We report a case of cutaneous metastatic undifferentiated pleomorphic sarcoma of the buttocks occurring in a 73-year-old man diagnosed with mediastinal sarcoma 4 years previously. He first noticed the mass approximately 2 months previously. Histological findings with immunomarkers led to a final diagnosis of cutaneous metastatic sarcoma from mediastinal undifferentiated pleomorphic sarcoma. **(Ann Dermatol 27(3) 310～314, 2015)**

-Keywords-
Malignant fibrous histiocytoma, Undifferentiated pleomorphic sarcoma

INTRODUCTION

Sarcomas can be classified as liposarcomas, fibrosarcomas, pleomorphic sarcomas, leiomyosarcomas, rhabdomyosarcomas, angiosarcomas, or osteosarcomas[1]. Undifferentiated pleomorphic sarcomas are a group of pleomorphic high-grade sarcomas that cannot be classified into any other subtype[1,2]. Undifferentiated pleomorphic sarcomas, known as malignant fibrous histiocytoma, is a malignant neoplasm that arises both in soft tissue and bones[1,3]. Histopathologically, it is described as an undifferentiated pleomorphic sarcoma characterized by atypical pleomorphic spindle tumor cells with a storiform growth pattern[3]. Undifferentiated pleomorphic sarcomas account for 5% of adult soft tissuesarcomas[4]. They predominantly present in the 5th to 7th decades of life and occur most frequently in the lower extremities[3]. Although undifferentiated pleomorphic sarcoma has high incidences of local recurrence and metastasis, to our knowledge, only 10 cases of cutaneous metastasis have been reported. Here, we report a case of cutaneous metastatic undifferentiated pleomorphic sarcoma on the right buttock area arising from a mediastinum sarcoma and review the relevant literature.

CASE REPORT

A 73-year-old man visited our department with a 3.0×3.5 cm deep seated nontender tumor on his buttock. He had a history of mediastinal sarcoma that had been excised 4 years previously. At that time, the tumor was a sarcoma with fibrotic and myxoid feature; it was excised completely, and the resection margin was clear. Thereafter, he was followed up by computed tomography (CT) without any recurrence. About 5 months previously, he complained of chest pain. CT showed a 12 cm mass in the left hemithorax with chest wall invasion and mediastinal mass excision was consequently performed. During follow-up, bone scintigraphy showed skeletal metastasis of the left 6th to 7th rib lateral arcs, and positron emission tomography CT showed reactive mediastinal lymph nodes.

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Therefore, the patient was treated with the following radiation schedule: X-rays, 180 cGy per fraction, and a total dose of 5,940 cGy. The patient was subsequently treated with 2 cycles of doxorubicin and ifosfamide. Meanwhile, a tumor was observed on the buttock and he was referred to us. On physical examination, the tumor on the right buttock appeared as a deep seated nodule (Fig. 1A, B). CT showed a 3×4 cm irregularly shaped soft tissue mass in the subcutaneous layer of his right buttock (Fig. 1C, D).

Histopathological examination of the tumor showed spindle-shaped cells arranged in a storiform pattern as well as atypical mitoses and pleomorphism. Immunohistochemistry revealed the tumor cells were positive for vimentin and Ki-67 (30%) but negative for CK7, CK19, EMA, S-100 protein, CD34, bcl-2, and CD99 (Fig. 2).

We diagnosed the tumor as cutaneous metastatic undifferentiated pleomorphic sarcoma. The patient was subsequently referred to the Department of General Surgery. The tumor was excised completely with a clear resection margin, and he was treated with chemotherapy using pazopanib.

DISCUSSION

The local recurrence rate of undifferentiated pleomorphic sarcomas is 19% to 31%, and the metastasis rate is 31% to 35% after tumor resection with a clear resection margin[5]. Despite the high distant metastasis rate, cutaneous metastasis is very rare.[6] The most common sites of metastasis are the lungs (90%), bone (8%), and liver (1%)[5,6]. Meanwhile, the cutaneous metastasis rate is 0.6% to 1.5%[7,8]. To our knowledge, only 10 cases of cutaneous metastasis have been reported (Table 1)[5-12]. Clinically, patients complain of a mass appearing rapidly, ranging from weeks to months[13]. Furthermore, the mass is usually asymptomatic[13]. In the present case, the patient did not complain of pain. Histologically, undifferentiated pleomorphic sarcoma is characterized by the presence of spokes of a wheel-like structure, showing spindle-shaped cells such as fibrocytes and histiocytes arranged in a storiform pattern[3,5,14]. Numerous giant cells show eosinophilic cytoplasm on staining; single or multiple irregular nuclei are observed, with inflammatory cells often mixing with the tumor cells[13]. In the present case, spindle-shaped cells were arranged in a storiform pattern and numerous pleomorphic cells with high mitotic rates were observed. Immunohistochemical staining for vimentin and CD68 is usually positive[14]. Moreover immunohistochemical staining for calretinin, cytokeratin, TTF-1, CD56, LCA, CD34, and S-100 proteinare negative[14]. However, there is no spe-

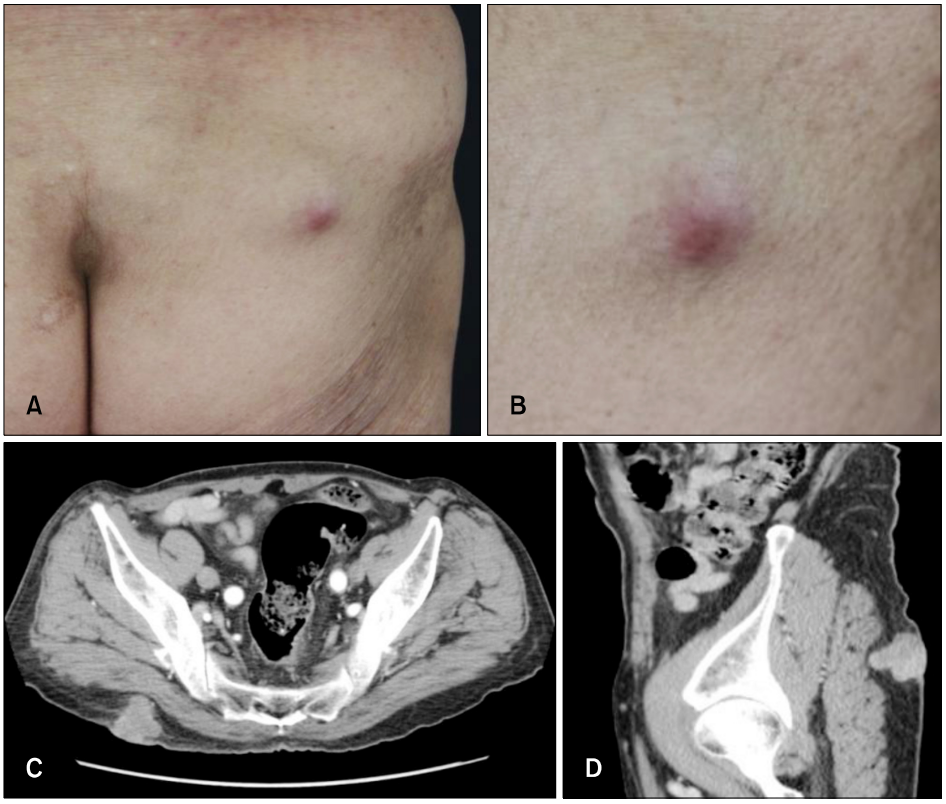

Fig. 1. (A) A solitary hard deep seated nodule in the right buttock. (B) Close-up of the nodule in (A). Axial (C) and sagittal (D) computed tomography images showing a 3×4 cm irregularly shaped metastatic soft tissue mass in the subcutaneous layer of the right buttock.

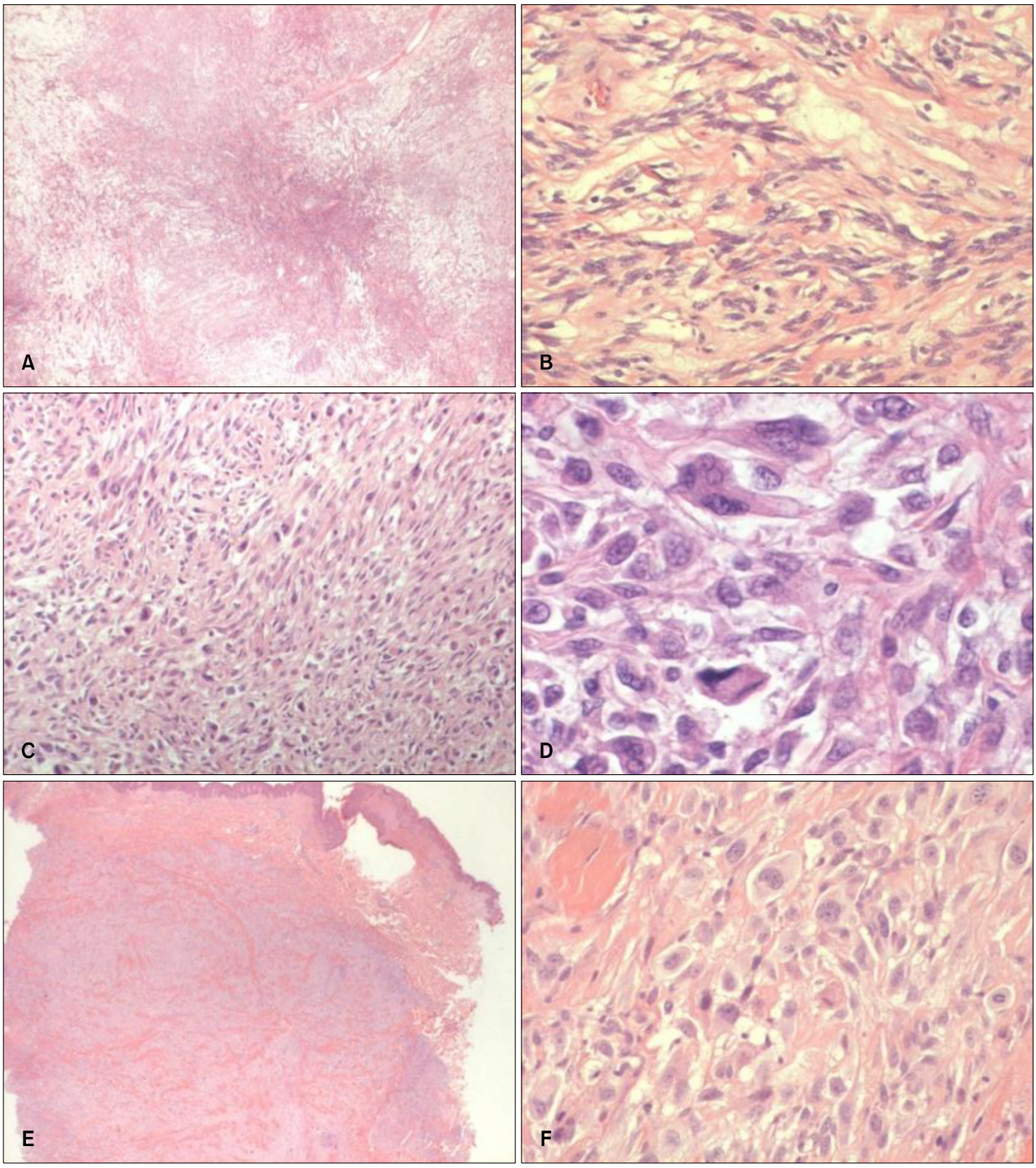

Fig. 2. The mediastinal tumor 4 years previously (H&E; A: ×12.5, B: ×200). The tumor was composed of spindle-shaped cells arranged in a storiform pattern with atypical mitoses the same findings were observed in the mediastinal tumor upon recurrence (H&E; C: ×12.5, D: ×400) and metastatic tumor in the buttock (H&E; E: ×12.5, F: ×400).

cific immunohistochemical stain that confirms the diagnosis[15]. The present case was positive for vimentin and Ki-67 (30%), and negative for CK7, CK19, EMA, S-100 protein, bcl-2, and CD99.

The differential diagnosis includes metaplastic (sarcomatoid) carcinoma, fibrosarcoma and other tumors such as malignant phyllodes tumor, inflammatory myofibroblastic tumor, and dermatofibrosarcoma[1,5]. Metaplastic (i.e., sarcomatoid) carcinoma, also called spindle cell carcinoma, is confirmed by CK positivity[1]. However, the present case,

Table 1. Summary of cutaneous metastatic undifferentiated pleomorphic sarcomas

Case No.	Author	Site of origin	Site of metastasis	Treatment	Survival after metastasis
1	Suzuki et al.[5]	Mandible	Skin, lung, liver, bone	NS	8 weeks
2	Lew et al.[6]	Breast	Ankle, liver	Excision	2 years (alive)
3～5	Weiss and Enzinger[7]	NS	Skin	NS	NS
6	Kim et al.[8]	NS	Skin, lung, brain, mediastinal LN, adrenal, abdominal LN, bone	Excision, chemotherapy	14 weeks
7	Enion et al.[9]	Upper arm	Thigh	Wide local excision	10 weeks
8	Kearney et al.[10]	NS	Scalp	NS	NS
9	Chen[11]	Retroperitoneum	Scalp, breast, liver, lung	Excision	NS
10	Geist et al.[12]	Lung, chest wall	Mandible, tongue	Chemotherapy, excision, radiation	NS
11	Present case	Mediastinum	Buttock	Excision	14 weeks (alive)

NS: not specified, LN: lymph node.

was negative for CK. Fibrosarcoma is another differential diagnosis but is less pleomorphic[5].

Cutaneous metastatic undifferentiated pleomorphic sarcoma is important because the prognosis can be very poor[5,6]. Among the 10 reported cases of cutaneous metastases, the survival period is only reported in 4; the average survival period in 3 patients was 8 to 14 weeks[5,8,9], whereas the other patient was alive for 2 years[6].

Interestingly, in the present case, the metastatic site was the buttock which is the most common intramuscular injection site. Although there was no evidence that the metastatic site was exactly the injection site, he complained of a mass after injection, followed by rapid tumor growth. Thus, it is possible the trauma caused by the injection was the trigger factor. In previous case reports, undifferentiated pleomorphic sarcoma was associated with burn, radiotherapy, and vaccination scars; a chronic ulcer at a fracture site; and a skin- graft donor site[16]. Thus, it is interesting that undifferentiated pleomorphic sarcoma occurred at an intramuscular injection site in the present case.

The treatment of choice for undifferentiated pleomorphic sarcoma is wide excision. Adjuvant radiotherapy is sometimes effective, and adjuvant chemotherapy can be performed in cases of multiple metastases[8]. In the present case, the tumor was completely excised with a clear resection margin, and chemotherapy with pazopanib was performed.

As cutaneous metastatic undifferentiated pleomorphic sarcoma is rare most clinicians do not have experience diagnosing this disease, which may result in a late diagnosis. Therefore, further study of this tumor is important, and the case reported herein expands knowledge about this tumor.

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