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A case of undifferentiated pleomorphic sarcoma of the breast with lung and bone metastases

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ABSTRACT

INTRODUCTION: Undifferentiated pleomorphic sarcoma (UPS) constitutes less than of all sarcomas in adults and rarely involves the breast. We herein present a patient with UPS of the breast with lung and bone metastases. This case was treated by eribulin as first chemotherapy, and performed mastectomy for local control.

CASE PRESENTATION: A 55-year-old female presented a tumor measuring over 5 cm with pain in the right breast. Pathology of the incisional biopsy specimen led to a diagnosis of UPS. Computed tomography revealed a right tumor, right pubic tumor with osteolysis, and multiple lung metastases. She was started on eribulin; however, the tumor grew in size, indicating progressive disease, and the patient underwent simple mastectomy for local control. Pathological evaluation of the excised tumor was consistent with UPS. The patient elected palliative treatment and died due to respiratory failure caused by multiple lung metastases that exacerbated four months after surgery.

DISCUSSION: Soft tissue sarcomas with distant metastases are treated with chemotherapy; however, there are currently no effective chemotherapeutic agents for UPS of the breast. Given the potential efficacy of eribulin in soft tissue tumors and the easy management of associated side effects, the patient was treated with eribulin, which however was insufficient for disease control.

CONCLUSION: The prognosis of UPS with distant metastasis remains poor. Treatment approaches including chemotherapy and surgery should be considered based on the patient's general condition, prognosis, and expectations on quality of life.

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1. Introduction

Primary breast sarcomas are extremely rare, representing less than 1% of all primary breast malignancies [1]. Prognosis of primary breast sarcomas is associated with tumor diameter, distant metastasis, and histological grade [2]. Lymphatic spread is uncom-

mon in primary breast sarcomas, and dissemination usually occurs hematogenously, and lungs, bones, and liver are the primary organs for metastasis [2]. Conversely, undifferentiated pleomorphic sarcoma (UPS) constitutes less than 5% of all sarcomas in adults and has been rarely reported to involve the breast [3]. There are no comprehensive reports on primary UPS of the breast, and individual case reports are rare. We herein present the case of a patient with suspicious UPS of the breast with lung and bone metastases.

The work has been done in line with the SCARE criteria [4].

2. Presentation of case

A 55-year-old female presented to a local clinic with a tumor measuring over 5 cm with pain in the right breast. Inflammatory granuloma was suspected, and the patient was followed up. However, the tumor grew in size, and an incisional biopsy was performed six months after the first visit. Pathological examination led

Abbreviations: CT, computed tomography; PD, progression disease; ER, estrogen receptor; PR, progesterone receptor; EMA, epithelial membrane antigen; SMA, smooth muscle actin; UPS/MFH, undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma; US, undifferentiated/unclassified sarcomas; NOS, not otherwise specified; QOL, quality of life.

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to a diagnosis of UPS, and the patient was admitted to our hospital for treatment.

The patient had no medical history, and her family history included breast cancer in her aunt. On physical examination, the patient had a tumor measuring 50 mm in her right breast, and her Eastern Cooperative Oncology group (ECOG) performance status (PS) was poor with a score of 3. Mammography showed a microlobulated tumor occupying the lower outer quadrant of the right breast. Ultrasonography showed a low-echoic mass with irregular margins and a heterogeneous internal echo pattern (Fig. 1). Computed tomography showed right breast tumor, tumor in right pubic bone with osteolysis, and multiple lung metastases (Fig. 2). The patient was diagnosed as UPS with distant metastases and initiated on eribulin mesylate, a nontaxane microtubule dynamics inhibitor, considering her PS score and tolerability of treatment. She received three courses of eribulin without significant side effects. However, the primary tumor was observed to grow noticeably on physical examination week 9 after treatment initiation, indicating progressive disease. Due to concerns regarding self-destruction of tumor and bleeding, simple mastectomy was performed for local control.

Pathological analysis revealed that the tumor which exhibited a milky appearance by gross examination appeared necrotic and bleeding internally. Microscopic assessment of the sections prepared from the surgical specimen showed spindle-shaped cells with heteromorphic strong nuclei (Fig. 3). Immunohistochemistry showed that the tumor was negative for estrogen receptor, progesterone receptor, AE1/AE3, CAM5.2, CD34, epithelial membrane antigen, and desmin and was partially positive for smooth muscle actin and S-100. These results were consistent with the diagnosis of UPS. After surgery, the patient chose palliative treatment after consultation and died due to respiratory failure caused by multiple lung metastases which exacerbated four months after the surgery.

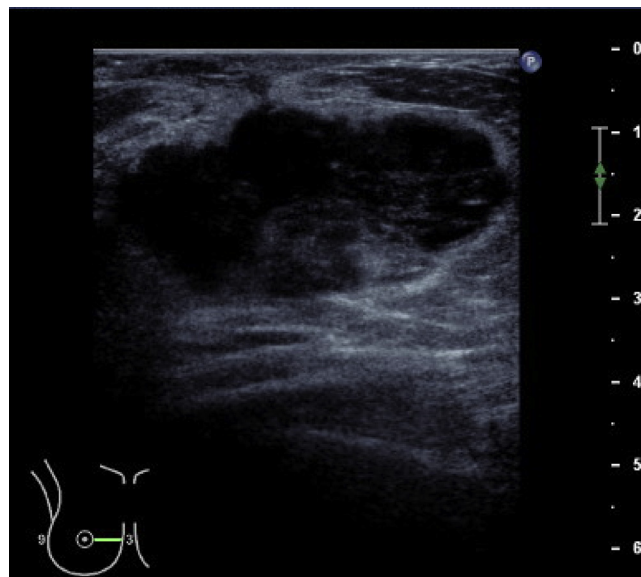


Fig. 1. Breast ultrasonography. Breast ultrasonography shows a low-echoic tumor measuring 51 mm in diameter with irregular margins and a heterogeneous internal echo pattern in the AB area of the right breast.

3. Discussion

In 2013, the WHO classification of soft tissue tumors was revised and the concept of undifferentiated pleomorphic sarcoma / malignant fibrous histiocytoma (UPS / MFH) disappeared, and a major item of undifferentiated / unclassified sarcomas (US) was created on behalf of UPS / MFH. In the definition of US, at most 20% of the whole soft tissue sarcoma is said to correspond to this disease concept [5]. There are five subtypes in

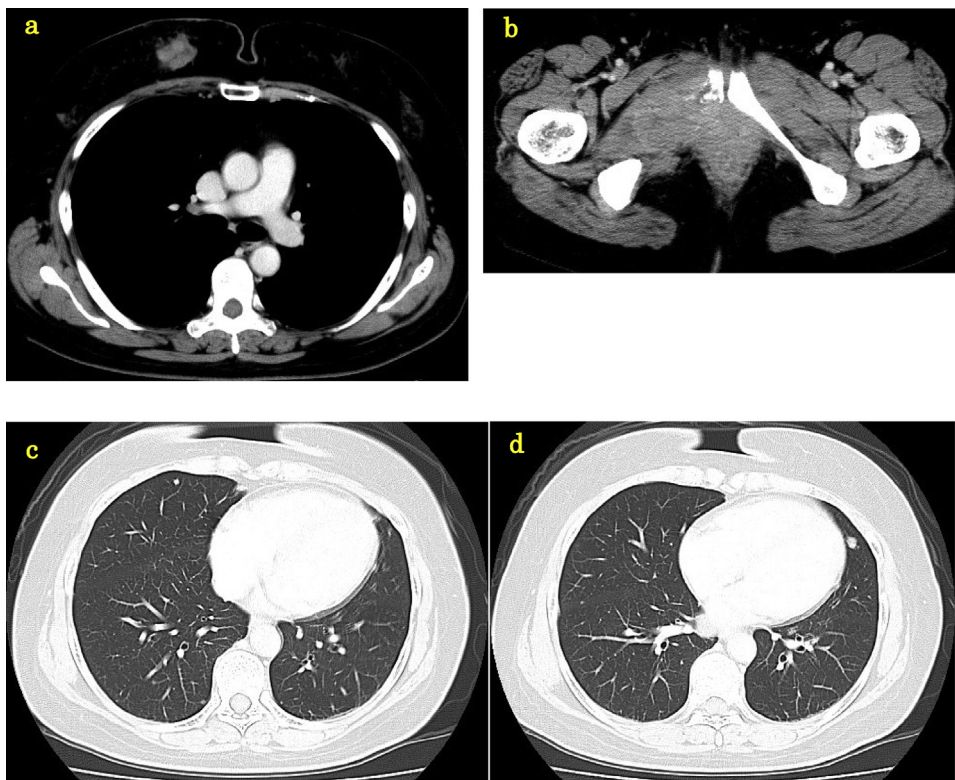


Fig. 2. Computed tomography. a. Computed tomography image showed right breast tumor. b. Computed tomography image showing right pubic tumor with osteolysis. c, d. Note the multiple lung metastases.

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