CASE REPORT

A 65-year-old Taiwanese female initially presented an 1-year history of experiencing an asymptomatic dark reddish subungual mass on the left thumb (Fig. 1A). The patient’s medical history included diabetes mellitus, hypertension, coronary arterial disease, and a right hemiparesis resulting from an intracerebral hemorrhage ten years prior. No history of trauma, irradiation, or chemical exposure to her hands was disclosed. A skin biopsy from the periphery of the lesion was performed and histopathologic examination revealed multinucleated giant cells and mononuclear inflammatory cells dispersed in the dermis (Fig. 1B, C). A diagnosis of giant cell tumor of tendon sheath was made to the patient. The patient preferred to preserve the function of the left thumb and undergo a conservative course of treatment so the tumor was removed by treatment with a carbon dioxide laser. At that point the patient discontinued further follow-up.

Three years later, this patient presented to our clinic with a singular, painful and gradually enlarging, ill-defined, pink-colored nodule at the same site on her left thumb (Fig. 2). Computed tomography (CT) detected a heterogeneous, well-enhancing soft tissue mass occupying the distal phalanx of the left thumb, along with osteolytic destruction of the distal phalanx bone. Histopathologic examination of a biopsy taken from the tumor revealed spindle cells forming a storiform pattern and focal areas composed of multinucleated giant cells. These tumor cells were immunoreactive to vimentin, but not S-100, cytokeratin, desmin, CD31, or CD34 (Fig. 3). The patient was subsequently diagnosed with malignant fibrous histiocytoma (MFH) of the storiform-pleomorphic type. For the purpose of curative treatment, amputation of the left thumb and partial resection of the proximal phalanx salvage was performed. The patient refused any additional survey for metastasis and discontinued follow-up after surgery.

DISCUSSION

MFH is the most common soft tissue sarcoma in people of 50 to 70 years old. Previous irradiation, or exposure to reagents such as phenoxy acid, are thought to be possible precipitating factors of the condition, although the precise etiology of MFH remains unknown. MFH typically involves the skeletal muscle or deep fascia of the proximal extremities, especially the thigh. Presentation of MFH in the digits is unusual, and there only are 6 published cases, including our patient.

MFH can be classified into four types based on histopathologic analysis: the storo
riform-pleomorphic type accounts for the majority, followed by the myxoid, giant cell, and inflammatory types. There are no definitive criteria for the diagnosis of MFH, and it is not differentiable from other types of sarcomas by clinical presentation or gross appearance. The diagnosis of MFH is based on the exclusion of other soft tissue sarcomas, immunohistochemical staining to demonstrate immunoreactivity to vimentin, and the absence of epithelial, endothelial, myogenic, neural, and melanocytic markers. In reviewing the medical history of our patient, the previous diagnosis of giant cell tumor of tendon sheath might have resulted from an inadequate biopsy site and a small specimen. If the specimen was taken from the periphery of the tumor, this would not represent the core tumor cell type. Giant cell tumor of tendon sheath and MFH are not similar, no matter clinically or histologically, and there is no evidence showing association between them in the literature. The clinical manifestation and histologic examination were not typical of MFH three years prior; we gave the diagnosis of giant cell tumor of tendon sheath according to the lesion site and histologic features. No immunohistochemical stain was obtained at that time, and this might obscure the diagnosis. The patient discontinued further follow-up, so it was impossible to observe the result of treatment or arrange further survey. Detailed pathologic studies, including necessary immunohistochemical stains, were performed three years later, and the diagnosis of MFH was made.

MFH has also been shown to be present following bone infarct and chronic osteomyelitis. The incidence of MFH in bone is 6%, with 36% of cases located in the femur. The tibia and humerus bones are also common sites for MFH, while the incidence for MFH in the phalanx bone is rare. When MFH is detected in both soft tissue and bone, the original MFH site may have been in the soft tissue followed by secondary extension into the bone, or vice versa. Time sequence studies using radiography can detect changes
Malignant Fibrous Histiocytoma in bone and local soft tissues which indicate a problem, while histopathologic examination can confirm the presence and origin of the tumor. In our patient, CT examination revealed a soft tissue mass accompanied by osteolytic destruction of the distal phalanx bone. It could not be determined whether the soft tissue lesion or compromise of the bone appeared first; however, histopathologic analysis indicated there was no evidence of the MFH originating in the distal phalanx bone.

Treatment of MFH usually involves wide local excision, although Mohs micrographic surgery is another treatment option. The infiltrative phenotype and poorly defined margin of MFH are the phenotypes associated with an aggressive tumor, so adjuvant chemotherapy or radiotherapy after surgery may provide added benefit. Local recurrence resulting from the spread along fascial planes or between muscle fibers is more often encountered in patients who are older than 50 years of age, or have MFH located in the upper extremities or distal sites. Distant metastases and local recurrences often occur within 12 to 24 months of diagnosis, and common metastatic locations include lung (90%), bone (8%), and liver (1%). In our patient, MFH recurred locally within three years, and the left thumb was disarticulated with partial proximal phalange salvage to obtain a wide excision. A chest radiograph showed no evidence of metastasis to the

Fig. 2
Imaging of the enlarged, poorly demarcated, pink-colored nodule with focal brownish hue located on the left thumb that developed three years after the initial nodule was examined.

Fig. 3
(A) The tumor cells infiltrating the lower dermis and subcutis. (H&E, original magnification x40)
(B) Spindle cells forming a whorled pattern with bizarre multinucleated giant cells scattered throughout the tumor. (H&E, original magnification x100)
(C) Staining of tumor cells reactive to vimentin. (Vimentin, original magnification x100)
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lung. Detailed blood biochemistry, abdomen sonography, and a bone scan were advised to the patient to evaluate possible metastases to the liver and bone; however, the patient refused further examination to survey local or metastatic spread and was lost to follow-up after surgery.

REFERENCES