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Malignant peripheral nerve sheath tumor of the distal phalanx of the fifth toe: a case report

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Abstract

Malignant peripheral nerve sheath tumor (MPNST) involving bone is rare. We report a case of MPNST of the fifth toe. The lesion was located in the distal phalanx of the right fifth toe and extended into surrounding subcutaneous tissues. Findings on magnetic resonance imaging and histological features of the case are described and the literature is briefly reviewed.

Keywords

Malignant peripheral nerve sheath tumor, distal phalanx, magnetic resonance imaging (MRI)

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Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a malignant soft tissue tumor arising from peripheral nerve sheath cells. MPNST is uncommon and can be associated with neurofibromatosis type I (NF-I). MPNST involving bone is very rare, and this is the first reported case of MPNST involving the distal phalanx of the fifth toe in a patient without NF-I.

Case report

A 76-year-old man with no personal or family history of NF-I presented with a 1-year history of a swollen, red, and slightly painful right fifth toe. He had consulted the Dermatology Clinic at Kobe University Hospital because of pain and occasional bleeding. He had a 7-year history of diabetes mellitus that was being treated with insulin. Examination revealed no signs of NF-1. The right fifth toe was swollen, indurated, and erythematous, with nail destruction and a small ulcer.

High-resolution magnetic resonance imaging (MRI) was performed using a 5-inch diameter microscopy coil, showing a tumor occupying the whole distal phalanx, with irregular thinning of the cortical bone and extension into subcutaneous soft tissues. The lesion was hypointense to muscle on T1-weighted (T1W) imaging and homogeneously hyperintense on T2-weighted (T2W) imaging and short-inversion-time inversion-recovery imaging. Heterogeneous enhancement was observed after gadolinium administration, mainly in the marginal regions (Fig. 1).

Histological examination of a skin biopsy specimen showed massive cell proliferation in a storiform pattern extending from the superficial dermis to the subcutis, predominantly comprising spindle-shaped clear cells with abundant cytoplasm and atypical

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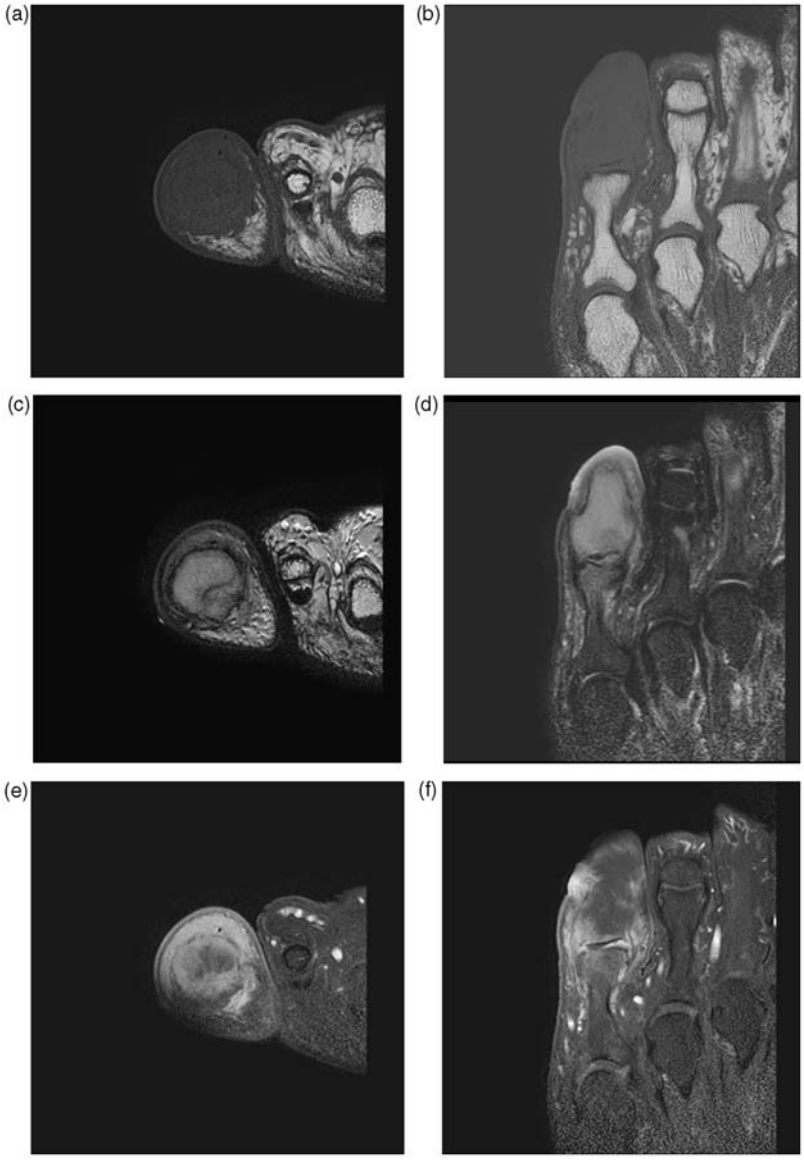

Fig. 1. (a, b) Axial and coronal T1W images showing a hypointense mass in the distal phalanx of the right fifth toe with widespread bone destruction. (c, d) T2W images showing a homogeneous hyperintense mass with cortical bone destruction and extension into subcutaneous soft tissues. (e, f) T1W images with gadolinium enhancement showing heterogeneous enhancement of the lesion.

hyperchromatic nuclei (Fig. 2a). Immunohistochemical staining for S-100 showed moderately positive results in the atypical spindle cells (Fig. 2b), whereas staining for desmin, α-smooth muscle actin, factor XIIIa, CD34, cytokeratin (AE1/AE3), HMB-45, and melanA yielded negative results (Fig. 2c). Malignant melanoma and clear cell sarcoma were therefore excluded as possible diagnoses. A diagnosis of MPNST involving the bone was made, and the right fifth toe and fifth metatarsal bone were amputated with no adjuvant chemotherapy.

At 1 year after surgery, local recurrence was detected, and further investigation revealed multiple pulmonary metastases.

Discussion

MPNST is a malignant neoplasm of the connective tissues and nerves. NF-I is the most significant risk factor for MPNST. Approximately 50% of MPNSTs develop in patients with NF-I, and 4.6% of NF-I patients develop MPNST (1). MPNSTs in non-NF1 patients occasionally arise from major nerves such as the sciatic nerve, sacral nerve, or brachial plexus.

The most frequent site of MPNST involving the bone is the mandible (2). It has been proposed that the long course of the alveolar nerve through the mandible may predispose to development of the tumor. However, this view is questioned, as other nerves with

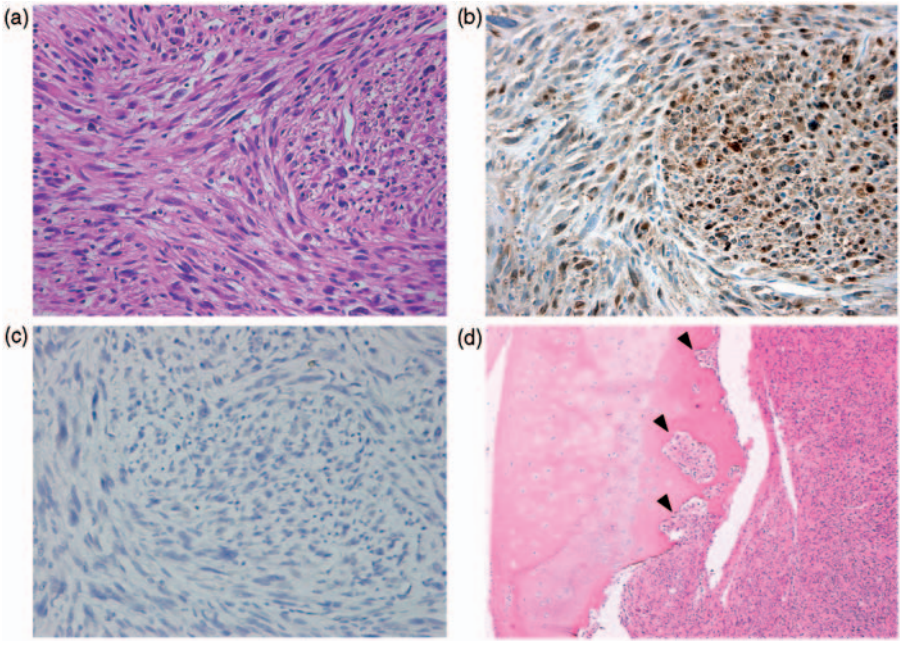

Fig. 2. (a) Cell proliferation in a storiform pattern, comprising mostly spindle-shaped cells with clear abundant cytoplasm and atypical hyperchromatic nuclei (hematoxylin and eosin stain, original magnification ×40). (b) Immunohistochemical staining for S-100 shows moderately positive results in tumor cells (original magnification ×40). (c) No expression of melanA is seen in tumor cells (original magnification ×40). (d) Typical spindle cell proliferation infiltrating skeletal tissues (black arrowheads) (hematoxylin and eosin stain, original magnification ×4).

extensive intraosseous courses show low frequencies of MPNST. Other sites of MPNST involving bone include the vertebrae (3), femur (4), and calcaneus (4). MPNST involving the bones of the digits is very rare. To the best of our knowledge, only one case of MPNST of the digits has been reported, in the distal phalanx of the thumb of a 6-year-old girl who did not have NF-I (4). The incidence of MPNST is considerably higher in non-NF1 children than in non-NF1 adults (5). The present case is unique in that the MPNST arose in the subcutaneous tissue of the fifth toe of a non-NF1 patient, where there are no major nerves.

MPNST involving bone is rare because MPNST usually arises in the soft tissues. There are three possible mechanisms for bone involvement in MPNST. First, the tumor may arise from within the bone, as intraosseous MPNST. Second, the tumor may invade the bone through a nutrient foramen, producing a dumbbell-shaped lesion as it enlarges. Third, an extraosseous tumor arising in the soft tissues may invade the adjacent bone (6,7). In this case, the bone was not involved via a nutrient foramen and there was no dumbbell shape to the lesion. The lesion involved both the bone marrow of the distal phalanx and the subcutaneous fat around the bone, and determining whether the tumor originated in bone or soft tissue was difficult. From the perspective of MRI findings, the greater bulk of the tumor was intraosseous, the bone marrow seemed to be totally infiltrated, and the lesion seemed to have arisen within the bone. However, based on the histopathological features, the tumor is thought to have originated in soft tissues (Fig. 2d) and involved the adjacent whole bone through penetration.

Most tumors arising in the digits are small, and there are technical limitations to the spatial resolution that can be achieved when evaluating such tumors using conventional MRI. In this case, high-resolution MRI using a 5-inch-diameter microscopy coil allowed us to evaluate the features and extent of the tumor. Heterogeneous enhancement of the marginal region reflected internal degeneration of the tumor (Fig. 1f). Use of a microscopy coil yields a high signal-to-noise ratio, and high-resolution MRI using a microscopy coil is therefore useful for evaluating small lesions of the hand and foot. Preoperative evaluation of the location and extent of the lesion are very important for surgical planning (8).

The imaging findings of MPNST involving bone are non-specific. Relative to muscle intensity, MRI usually shows an isointense lesion on T1W imaging and a hyperintense lesion on T2W imaging, with various patterns of enhancement depending on the degree of degeneration (3,9), as in our patient. Cortical thinning

of the bone and invasion of the soft tissues around the bone are usually accepted as signs of malignancy, but have also been described in benign schwannoma (10).

Differential diagnoses for the lesion in our patient included osteolytic lesions such as bone metastasis, lymphoma, plasmacytoma, and malignant fibrous histiocytoma, and soft tissue tumors such as metastasis, synovial sarcoma, and dermatofibrosarcoma protuberans.

In conclusion, diagnosis of MPNST with bone involvement may be difficult in small bones because of the rarity of the condition and the non-specific radiological findings, and definitive diagnosis requires histopathological examination.

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