Osteosarcoma of the middle and distal phalanges of the little toe with a cancerous ulcer

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ABSTRACT

Introduction: Osteosarcoma (OS) is the most common malignant mesenchymal tumor of the bone, and mostly affects the long bones around the knee. Osteosarcoma of short tubular bones such as those of the foot is extremely rare and accounts for less than 1% of all cases. To the best of our knowledge, this is the first reported case of OS of the middle and distal phalanges of the little toe.

Case Report: A 20-year-old Japanese male presented at our hospital due to pain and swelling of his left little toe for over one year. Physical examination revealed a swollen little toe on his left foot with skin ulceration and degenerative nail. Various radiological imaging tests including X-ray, CT scan, Magnetic resonance imaging (MRI) scan, positron emission tomography (PET) scan with 18F-FDG and bone scintigraphy with 99mTc -MDP showed tumors in the distal and middle phalanges of the left little toe with extraosseous masses and without distant metastasis. Open biopsy showed pleomorphic osteogenic sarcoma. After disarticulation at the metatarsophalangeal joint of the little toe, the patient received adjuvant chemotherapy with high-dose methotrexate, doxorubicin, and cisplatin. No recurrence has been observed on 12-month follow-up.

Conclusion: Pleomorphic osteogenic sarcoma is rare in the feet so the initial diagnosis is often wrong, leading to delayed appropriate treatment. This case report emphasizes the need to add osteosarcoma in the differential diagnosis of the phalangeal bone tumors.

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CASE REPORT

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Keywords: Foot phalanges, Osteosarcoma, Short tubular bones, Skin ulcer

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INTRODUCTION

Osteosarcoma (OS) is the most common malignant sarcoma of the bone, and involves the production of osteoid matrix by tumor cells. Osteosarcoma is typically seen in the metaphysis of long bones. In contrast, OS of short tubular bones such as those of the hands and feet is uncommon [1]. While OS in these locations has a favorable prognosis compared to conventional OS of the long bones, its initial diagnosis is often wrong due to its rarity, leading to delayed appropriate treatment [1, 2]. Since high-grade OS of the feet may be lethal, accurate diagnosis is critical. We present here a rare case of osteosarcoma of the little toe. To our knowledge, this is the first report of OS in this location that provides detailed radiological findings.

CASE REPORT

A 20-year-old Japanese male presented at our hospital due to pain and swelling of his left little toe for over one year. The patient had a history of trauma in the same toe two years previously. Examination revealed a swollen little toe on his left foot with skin ulceration. The nail showed degenerative changes (Figure 1). Family history and past medical history were unremarkable. Laboratory data, including serum total alkaline phosphatase and C-reactive protein, were within the upper limits of normal. Initial radiographic examination of the feet showed mixed calcification/ossification and osteolytic lesions with poorly defined margins and no apparent periosteal reaction in the distal and middle phalanges of the left little toe (Figure 2). X-ray examination also demonstrated fusion of the distal and middle phalanges of the right little toe, indicating that the fusion could be a normal variant (Figure 2, inset). On CT scan, popcorn-like calcification/ossification was clearly visualized with heterogeneous radiographic density (data not shown). Magnetic resonance imaging (MRI) scan showed tumors involving both the distal and middle phalanges with extraosseous masses, as well as disappearance of the subcutaneous fat (Figure 3). The tumor had a low-intermediate signal on T1-weighted image (T1WI), and a mixed low-to-high signal on SPAIR fat suppression. It demonstrated markedly heterogeneous Gd-contrast enhancement in both intraosseous and extraosseous portions. Both positron emission tomography (PET) scan with ^{18}F -FDG and bone scintigraphy with $^{99\text{m}}\text{Tc}$ -MDP depicted uptake (SUVmax 4.0 in PET) in the little toe corresponding to the primary site (Figure 4). Open biopsy showed pleomorphic sarcoma without apparent osteoid. Immunohistochemically, tumor cells were positive for SMARCB1/INI1 (nuclear), but negative for CAM5.2, AE1/AE3, desmin, alpha-SMA, beta-catenin, CD68, S-100 protein, and FGF23, which suggested pleomorphic sarcoma (data not shown). Although osteoid was not evident in the biopsy specimen, clinical, radiological, and pathological features led to a diagnosis

of OS of the middle and distal phalanges of the left little toe without distant metastasis. Since the tumors did not invade the proximal phalanx of the toe, disarticulation was performed at the metatarsophalangeal joint of the little toe. The amputated specimen was examined pathologically. As was the case with open biopsy, microscopy showed a proliferation of oval to polygonal tumor cells, with vesicular pleomorphic nuclei arranged in haphazard pattern, accompanied by irregular osteoid formation, staghorn vessels, and multinucleated tumor giant cells, which led to a pathological diagnosis of typical osteoblastic OS (Figure 5).

The patient's clinical course after the disarticulation of the little toe was uneventful. The patient received adjuvant chemotherapy with high-dose methotrexate, doxorubicin, and cisplatin. No recurrence has been observed on 12-month follow-up.

Figure 1: Local findings of the little toe on admission. The patient's left little toe was getting more swollen with skin ulceration.

Figure 2: (A) An anterior-posterior radiographs of both feet and (B) an oblique radiograph of the left foot; both (A) and (B) showing a mixed osteoblastic and osteolytic lesion in the left middle and distal phalanges of the little toe. Inset: oblique radiograph showing fusion of the right distal and middle phalanges of the little toe. Periosteal reaction is not evident.

DISCUSSION

Osteosarcoma (OS) is a rare malignant bone-forming mesenchymal tumor that accounts for 0.2% of all malignant tumors. In this report, we described a 20-year-old Japanese male who presented with OS in the little toe.

The incidence of OS of the hands and feet has been reported to be low, less than 1% of all cases [1]. Of reported OS cases of the feet, only a few occurred in the

Figure 3: MRI scan images of left little toe. T1WI (A–C), SPAIR fat suppression (D–F), and post-Gd (G–I) in the axial, coronal, and sagittal planes. The tumor showing a low-intermediate signal on T1WI and a mixed low-to-high signal on SPAIR, and arises from the middle and distal phalanges of the little toe and invaded the subcutaneous fat. The tumor showing markedly heterogeneous Gd-contrast enhancement in both intraosseous and extraosseous portions.

Figure 4: (A) Coronal PET images showing hypermetabolic focuses in the distal fifth toe. Maximum SUV was 4.0, and (B) Technetium-99-MDP bone scan showing avid uptake in the distal fifth toe (arrow).

Figure 5: (A) Macroscopic appearance of a formalin-fixed, cut section of the amputated left little toe, and (B) Histological specimen showing proliferation of oval to polygonal tumor cells, with vesicular pleomorphic nuclei arranged in haphazard pattern, accompanied by irregular osteoid formation (H&E stain, x100).

foot phalanges [2, 3]. Osteosarcoma of the proximal phalanx of the great toe was first reported by Mirra et al. in 1988 [2]. Recently, Anninga et al. found that 27 of 4221 cases of OS occurred in the foot [1]. They showed that clinicopathologically, patients with OS of the feet tended to be older, male, and with a lower grade of malignancy. As for tumor site, only 3 of 27 cases of OS occurred in the phalanges, 2 in the proximal phalanx of the great toe and one in the proximal phalanx of the second toe. According to the Bone Tissue Tumor Registry reported by the Musculoskeletal Tumor Committee of the Japanese Orthopedic Association, 4337 cases of OS were registered in Japan from 1972 to 2011 [4]. Twenty-nine cases (0.67%) were identified in the foot and only one occurred in a phalanx (the specific site was not documented). To our knowledge, no OS cases in the little toe phalanges have been previously described.

Osteosarcoma of the feet is extremely rare so the initial diagnosis is often wrong, leading to delayed appropriate treatment. In the present case, at the patient's initial visit the macroscopic appearance of the lesion was similar to that of skin tumor or infectious granuloma due to skin ulceration and granulation. Histological examination in the biopsy specimen showed pleomorphic sarcoma without evident osteoid, various imaging studies and the histological features made us a clinical diagnosis as OS.

Radiographic features of OS of the feet have not been well documented. In the present case, plain X-ray and CT scan demonstrated mixed osteoblastic and osteolytic lesions of the distal and middle phalanges of the little toe, which are typical radiographic findings of long bone OS. Periosteal reaction such as Codman's triangle and sun-burst appearance is another typical radiographic finding of long bone OS, but it was not observed in the present case. It is reported that OS associated with craniomaxillofacial fibrous dysplasia frequently lack a periosteal reaction [5]. Thus, we speculate that periosteal reactions in OS might be site-dependent.

On MRI examination, OS as well as other malignant bone tumors mostly exhibit a low-intermediate signal on T1WI and a low-high signal on T2WI, with or without fat suppression, and therefore these findings are non-specific

for OS [6], as observed in our case. Primary malignant bone sarcomas such as OS and Ewing sarcoma are usually accompanied by extraosseous lesions [7]. Although OS of the foot phalange is extremely rare, taken together with X-ray and CT findings, bone-forming tumor of foot phalanges with an intramedullary and extraosseous mass on MRI strongly suggests OS.

PET-CT using 18F-fluorodeoxyglucose (FDG) has been used for screening, staging, and evaluation of various cancers prior to neoadjuvant chemotherapy. Aoki et al. reported that OS demonstrated a relatively high SUVmax ($n = 6$; SUV, 3.07 ± 0.96) [8], and our patient had high SUVmax (4.0) as well. Therefore, FDG-PET-CT scan has become an important imaging modality for screening, staging and evaluation of OS before starting OS treatment. To the best of our knowledge, this is the first report describing PET-CT in OS of a foot phalanx.

In general, the treatment of high-grade OS consists of neoadjuvant chemotherapy, radical surgery, and adjuvant chemotherapy [9]. Although foot OS seems to have a favorable prognosis, Anninga et al. reported that these cases can be fatal and they therefore recommend treating high-grade OS of the feet similarly to that in conventional sites [1]. Our patient experienced severe pain in the little toe, and he could not put on his shoes. He therefore preferred immediate surgery to neoadjuvant chemotherapy, and we performed metacarpophalangeal disarticulation of the little toe in first. A large clinical trial showed that neoadjuvant chemotherapy failed to demonstrate survival superiority compared with adjuvant chemotherapy [10]. Since pathological diagnosis about the amputated specimen was typical osteoblastic OS, we performed adjuvant chemotherapy after the disarticulation.

CONCLUSION

This study is the first to report osteosarcoma (OS) of the little toe. Osteosarcoma of the feet is extremely rare, but can be lethal. Clinicians should consider the diagnosis of OS even when plain X-ray shows a mixed osteoblastic and osteolytic lesion with no periosteal reaction. Prompt biopsy and histological examination should shorten delays in diagnosis and improve outcomes of patients with OS of the feet.

Author Contributions

Tomoya Matsunobu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Hirofumi Bekki – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Katsumi Harimaya – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Yoshihiro Matsumoto – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Makoto Endo – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Kojiro Yoshitake – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Yoshinao Oda – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Yukihide Iwamoto – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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