Desmoplastic Fibroma Arising in the Distal Phalanx of the Great Toe: A Case Report

Takeshi Koba, MD1, Michiro Susa, MD, PhD1, Robert Nakayama, MD, PhD1, Itsuo Watanabe, MD1, Keisuke Horiuchi, MD, PhD1, Makio Mukai, MD, PhD2, Hiroo Yabe, MD, PhD1, Yoshiaki Toyama, MD, PhD1, Hideo Morioka, MD, PhD1

1Department of Orthopaedic Surgery, Keio University School of Medicine, Shinjuku-ku, Tokyo, Japan
2Department of Diagnostic Pathology, Keio University School of Medicine, Shinjuku-ku, Tokyo, Japan

ARTICLE INFO
Level of Clinical Evidence: 4
Keywords: adolescent bone desmoid frozen section hallux osteolysis spindle cells tumor

ABSTRACT
Desmoplastic fibroma (DF) of the bone is a rare locally aggressive tumor usually occurring in adolescents and young adults. These tumors most commonly occur in the mandibles and metaphyses of long bones but are extremely rare in small bones, often resulting in diagnostic problems. The occurrence of these tumors in the foot is especially limited. We report the clinical, radiographic, and histologic features of DF arising in the distal phalanx of the great toe and a review of the published data.

© 2014 by the American College of Foot and Ankle Surgeons. All rights reserved.

Desmoplastic fibroma (DF) of the bone was first described by Jaffe et al (1) in 1958. It is a rare entity reported to occur in approximately 0.1% of all bone tumors. It has a histologic resemblance to desmoid-type fibromatosis with similar aggressiveness locally. They usually present as a slow growing mass, most frequently occurring in the mandibles and long bones. Surgical resection with a wide margin has been advocated for the treatment. Curettage has been associated with a local recurrence rate as great as 50%, and, because of its rarity, data on the effect of adjuvant treatments on surgery are scarce. In 1986, el-Tabbakh and Al-Arabi (2) first described a case of intraosseous DF of a digit of the foot, and to our knowledge, the present case is the third such case occurring in the toe. We present a case of DF arising in the toe, a rare presentation with limited cases reported in published studies.

Case Report

An 18-year-old male presented with pain and a slow growing mass of his left great toe (hallux). He had no particular incidence of trauma or any relevant medical history. He had visited a nearby hospital because of deteriorating pain and was diagnosed with a bone tumor of the distal phalanx of the great toe radiographically. The overlying skin was smooth and nonadherent, with slight tenderness. No redness or hotness was present in the area. The blood examination results were all in the normal range. On the plain radiograph, a 2 × 3-cm osteolytic lesion was found in the distal phalanx with ballooning and thinning of the cortex. No periosteal reaction or calcification was associated with the lesion (Fig. 1). Because the patient refused to undergo surgery under general anesthesia, the operative plan was to perform an intraoperative biopsy of the lesion with the patient under local anesthesia and to perform extended curettage with bone grafting using allografts if the lesion was benign. The tumor was a white elastic hard mass with slight adhesion to the bone. On frozen section analysis, the tumor was composed of bundles of spindle cells with slight atypia and no mitosis, reminiscent of nonosseous fibroma. The operation was completed as planned, and the patient was released without any fixation (Fig. 2). On histopathologic examination after surgery, hematoxylin and eosin staining demonstrated dense bundles of spindle cells without the atypia and mitosis seen during surgery (Fig. 3). The immunohistochemical examination finding was negative for smooth muscle actin, desmin, S-100, and CD34. The MIB-1 index was less than 5%, consistent with DF. The postoperative period was uneventful, and, at the final follow-up visit after 3 years, no local recurrence, arthritic changes, or metastasis was observed (Fig. 4).

Discussion

DF of the bone is a benign, but locally aggressive, fibrous tumor usually occurring in patients younger than 30 years. The tumor was...
Fig. 1. (A) Anteroposterior and (B) oblique radiographs of the great toe at the initial presentation. The tumor is depicted as an osteolytic lesion with ballooning and thinning of the cortex.

Fig. 2. (A and B) Radiographs after surgery showing allograft grafted into the cavity.
first reported by Jaffe (1) in 1958, and Unni and Inwards (3) later reported that of the 10,165 primary bone tumors, 16 were DF (0.16%). The tumor occurs mostly in the jaw and metaphysis of the long bones, with a slight prevalence in males. Bohm et al (4) analyzed 184 cases of DF of the bone and reported that 23% resided in the mandible, 15% in the femur, 13% in the pelvis, 12% in the radius, and 9% in the tibia. To date, 10 cases of DF have occurred in the foot (Table), and the present case is the third occurring in the digit (2,5).

Clinically, the tumor is a slow growing, firm mass with increasing pain, ranging in duration from a few months to several years, with an occasional association with trauma. Pathologic fracture is uncommon. Radiographically, the lesion will be depicted as a well-delineated, lucent, and expansile mass with no evidence of periosteal reaction or mineralization. Although we did not perform computed tomography or magnetic resonance imaging, when the tumor is associated with cortex destruction, it is useful to assess its soft tissue extension. Magnetic resonance imaging studies are scarce, but reports have shown low to intermediate signal intensity on both T1-weighted and T2-weighted images. Histologically, the tumor will be analogous to its counterpart that occurs in the soft tissue, which is composed of spindle cells and various amounts of collagen fibers with scant mitosis. No giant cells or hemosiderin deposits will be observed in the lesion. Radiographically, it is sometimes difficult to distinguish from other tumors such as chondromyxoid fibroma, giant cell tumor, nonosseous fibroma, fibrous dysplasia, and bone cysts. The most important differentiation should be made between malignant tumors such as fibrosarcoma and low-grade osteosarcoma, for which a delay in diagnosis could lead to death. Because the radiographic characteristics of DF are quite nonspecific, a histologic examination is imperative. Fibrosarcoma of bone is differentiated by the presence of...
fibroblasts with atypia and mitotic figures. Low-grade osteosarcoma will contain osteoids.

Reports of genomic alterations in DF have been limited. There are conflicting reports of whether there is abnormality in the karyotypes. Trombetta et al (6) reported on recurrent chromosome aberration in 11q and 19p after short-term culturing of the tumor. Min et al (7) performed comparative genomic hybridization and identified multiple losses and gains in DF with malignant transformation; however, it is still difficult to establish the prognostic indicators for DF of the bone until additional studies are available. Hauben et al (8) analyzed 6 cases of DF, focusing on mutations in the β-catenin (CTNNB1) gene, all of which were negative. Although the histologic appearance of DF of the bone is quite similar to desmoid-type fibromatosis, the genomic background suggests that these 2 entities do not overlap.

Treatment of DF of the bone is still controversial because of its rareness. The tumor is locally aggressive, and local recurrence has been reported with high frequency. Gebhardt et al (9) reported that 42% of the tumors recurred after curettage but only 25% recurred after en bloc resection. Conflicting findings have been reported of surgical treatment of these rare entities, with some investigators recommending marginal to wide resection of tumors occurring in large bones and curettage for small lesions and others advocating wide resection for all lesions. Two cases of local recurrences in the foot have been reported, and both cases were treated with amputation with good results (10,11). Sporadic reports of radiotherapy have been reported for the treatment of DF (12–14); however, additional studies are needed to analyze its effectiveness as a definitive treatment. No extensive study has been published on adjuvant treatment during surgery. In the present case, heat ablation was performed using a standard electrosurgical knife after thorough curettage. Although the follow-up period was limited, no local recurrence has developed in the present patient, and limb salvage, instead of amputation, might be the treatment of choice for the initial treatment of these aggressive tumors occurring in digits.

Strict follow-up is imperative because of the tumor’s aggressive nature. No clear-cut consensus has been reached regarding the duration of follow-up that would be sufficient. Bohm et al (4) reported that of the 184 cases of DF, 28 cases recurred within 5 years. Desmoid-type fibromatosis, considered a counterpart in the soft tissue, also shows local aggressiveness and, studies have suggested that most of the recurrences develop within 3 years (15). Although sporadic cases of malignant transformation of DF after more than 10 years have been reported (14,16), they have been the exception, and patients with DF of bone should be followed up intensively for at least 3 years.

In conclusion, we have reported a rare case of DF of the distal phalanx of the great toe. Curettage and ablation was performed, and at 3 years postoperatively, our patient had no evidence of recurrence and was free of symptoms. DF of the bone should be considered in the differential diagnosis of tumors occurring in the foot. The rarity of this tumor has not enabled prediction of the possible outcomes after extended curettage with heat ablation. Because the rate of local recurrence is high, additional follow-up is warranted.

References