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Rapidly growing fibro-osseous pseudotumor of the digits mimicking extraskeletal osteosarcoma

Jun Nishio[1,2], Hiroshi Iwasaki[1], Osamu Soejima[2], Masatoshi Naito[2], and Masahiro Kikuchi[1]

[1] Department of Pathology, School of Medicine, Fukuoka University, 7-45-1 Nanakuma, Jonan-ku, Fukuoka 814-0180, Japan
[2] Department of Orthopaedic Surgery, School of Medicine, Fukuoka University, Fukuoka, Japan

Abstract Fibro-osseous pseudotumor of the digits is a rare benign lesion characterized histopathologically by a fibroblastic proliferation with foci of osseous differentiation. We report a case of fibro-osseous pseudotumor of the digits with a rapidly growing lesion in a 30-year-old woman. Because of its rapid growth and aggressive histopathological appearance, this lesion can be mistaken for a malignant neoplasm such as extraskeletal or parosteal osteosarcoma, despite its benign clinical behavior. For a soft tissue mass in the digit with a clinicopathological feature of pseudomalignancy, the clinician should consider fibro-osseous pseudotumor of the digits as a possible diagnosis, thereby avoiding inappropriately aggressive surgery.

Key words Fibro-osseous pseudotumor of the digits · Extraskeletal osteosarcoma · Bizarre parosteal osteochondromatous proliferation (Nora's lesion)

Introduction

Fibro-osseous pseudotumor of the digits is a benign lesion proposed as a unifying term by Dupree and Enzinger.[3] Typically, this lesion affects young adults, mainly women, and presents as a painful, localized, fusiform, and often erythematous swelling in the soft tissues of the fingers, especially the region of the proximal phalange. Fibro-osseous pseudotumor of the digits can be confused with a malignant neoplasm. In fact, there are several reports of cases in which the lesion was initially incorrectly diagnosed and the patient was treated by unnecessary radical surgery.[1,3,5] In the present study, we report a case of fibro-osseous pseudotumor of the digits that developed rapidly over a period of 4 weeks in a 30-year-old woman, and its clinicopathological features are described.

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Case report

A 30-year-old, right-hand-dominant woman presented with a mildly painful swelling in the palmar aspect of the middle phalange of the left ring finger (Fig. 1a). There were no local signs of infection and no constitutional symptoms such as fever, weight loss, or malaise. Laboratory studies revealed a normal white blood cell count of $6.7 \times 10^3/\mu l$, with 64.8% neutrophils and 29.5% lymphocytes. The rest of the laboratory examination results, including assessments of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), were within the normal limits. Four weeks later, the size of the swelling increased rapidly, and a firm mass of 2.0×1.8 cm appeared over the palmar aspect of the middle phalange of the left ring finger (Fig. 1b). Movement of the proximal interphalangeal joint of the left ring finger became restricted. Neural and vascular examinations were normal. There was no history of previous trauma, but the patient reported undergoing excision of a mass in the same region, 2 years earlier. The histological features of the excised lesion were suggestive of benign fibrous tumor (Fig. 2). However, no definite diagnosis could be made because of insufficient sampling.

A plain radiograph taken on admission revealed a mild amount of soft tissue swelling with calcification but without periosteal reaction (Fig. 3a). Four weeks later, a plain radiograph showed a severe degree of soft tissue swelling and calcification was twice as large as that noted on the initial radiograph (Fig. 3b). There was no periosteal reaction or bony erosion.

A local excision was performed without complications. The lesion showed little adherence to the surrounding soft tissues and had not invaded the flexor tendon or periosteum. The cut surface was well circumscribed, rubbery, and gray-white in color, measuring 2.3×1.8 cm. Light microscopy showed that the lesion was composed of a proliferation of typical

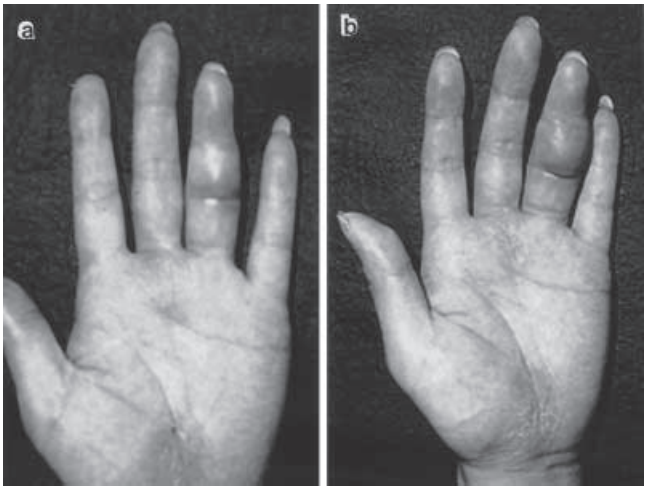

Fig. 1. a Initial palmar view of left hand showing a mild swelling of the ring finger. **b** Note the lesion on the ring finger and a rapid growth within a period of 4 weeks

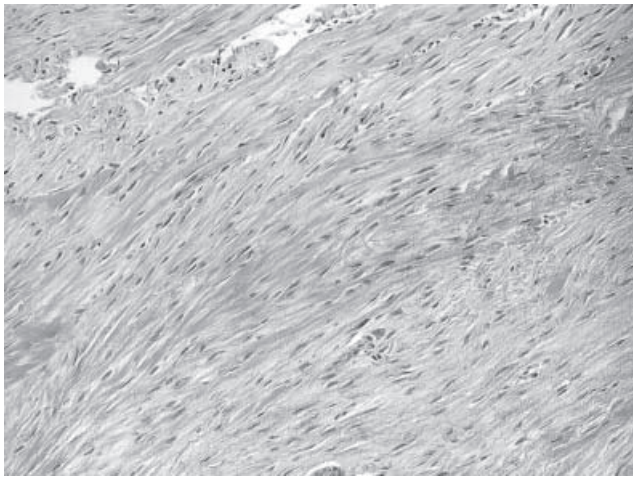

Fig. 2. A benign fibrous tumor preexisting at the site of the present tumor. There is a proliferation of fibroblasts in a collagenous matrix. Hematoxylin and eosin. ×100

fibroblasts admixed with foci of osteoids and bone trabeculae with no evidence of cellular atypia (Fig. 4a). The osseous trabeculae were distributed haphazardly with incomplete zoning phenomenon. In some areas, the fibroblasts were loosely dispersed within a prominent myxoid matrix (Fig. 4b). Abnormal mitotic figures and nuclear pleomorphism were absent. Immunohistochemically, the fibroblastic cells stained positive for vimentin, but were uniformly negative for AE1/AE3, CAM 5.2, EMA, desmin, HHF35, or S-100 protein. A small number of fibroblasts were weakly positive for alpha-smooth muscle actin. Based on these features, the lesion was diagnosed as fibro-osseous pseudotumor of the digits. No further treatment was necessary. There are no signs or symptoms of recurrence 7 months after surgery, and the patient has returned to her premorbid functional status.

Discussion

The recommended treatment of fibro-osseous pseudotumor of the digits is local excision. Unfortunately, however, the lesion can be misdiagnosed both radiographically and histopathologically as a malignant neoplasm. In fact, the correct clinical diagnosis was not given in two cases, and needless amputation of the digit was performed.[1,5] In addition, Dupree and Enzinger[3] reported six cases misdiagnosed as extraskeletal or parosteal osteosarcoma. Two patients were treated by amputation of the digit, one patient by radical resection before histological sections were reviewed. Therefore, it is of paramount importance to clearly distinguish this lesion from a malignant neoplasm, especially extraskeletal osteosarcoma. On the other hand, the rapid growth rate in our case led to a strong clinical suspicion of extraskeletal osteosarcoma. Although the distinction between these entities at an early stage may be difficult, a few distinguishing features may be useful in differential diagnosis.[3,6] (1) Extraskeletal osteosarcoma usually occurs in areas in the lower extremity, and involvement of the digits is rare. (2) Extraskeletal osteosarcoma is rarely encountered in patients younger than 35 years of age, whereas fibro-osseous pseudotumor of the digits prevails in young adults between the ages of 20 and 30 years. (3) Histopathologically, pleomorphism and atypical mitoses are seen in extraskeletal osteosarcoma, whereas no such features are seen in fibro-osseous pseudotumor of the digits. The distinction between the two entities should be finally made histopathologically as in our case.

The differential diagnosis may also include bizarre parosteal osteochondromatous proliferation (Nora's lesion). Given the overlapping clinical and histological features, it has been proposed that Nora's lesion most likely represents an intermediate step between fibro-osseous pseudotumor of the digits and acquired osteochondroma.[2] However, our lesion was not attached to the bone surface, and no definite cartilage cap or chondrocytes were observed. Thus, the diagnosis of Nora's lesion was not appropriate in our case.

The etiology of fibro-osseous pseudotumor of the digits is still unclear. In the Dupree and Enzinger series,[3] a history of trauma was given in only 9 of 21 patients, and both a neglected minor injury and infectious process have to be considered as a possible cause. There was no history of associated trauma or infection in our case. However, interestingly, 2 years

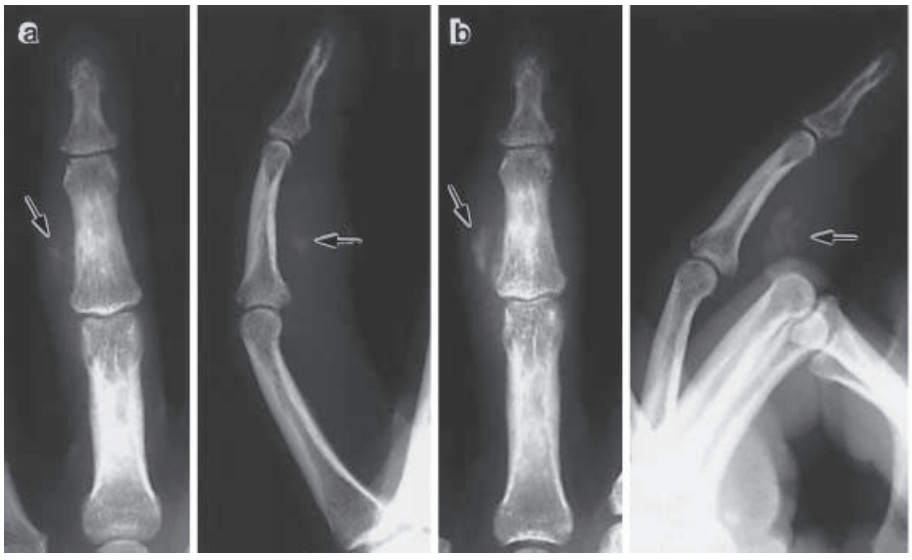

Fig. 3. a On admission, a plain radiograph of the left ring finger showed a mild degree of soft tissue swelling with calcification (*arrows*). There is no evident periosteal reaction. **b** Calcification (*arrows*) doubled in size radiographically 4 weeks after the onset of the lesion. There is no involvement of the underlying bone

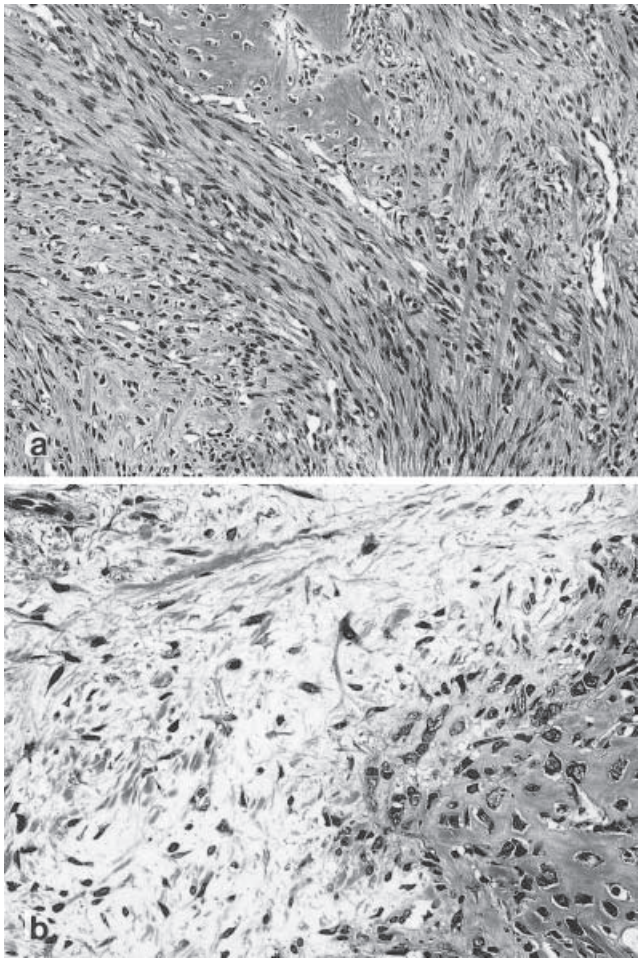

Fig. 4. a Cellular portion of a fibro-osseous pseudotumor of the digits composed of proliferated fibroblasts and focal deposits of osteoid. Hematoxylin and eosin. ×100. **b** Fibro-osseous pseudotumor of the digits shows a mixture of loosely arranged fibroblasts, a prominent myxoid matrix, and osteoids rimmed by uniform osteoblasts. Mitotic figures and nuclear pleomorphism are absent. ×160

earlier, our patient underwent resection of a mass in the same region. Thus, we cannot rule out the possibility that a trauma due to previous surgery plays a role in the development of the present lesion. Conversely, if the previous lesion was partly excised at an early phase of its growth, it is also possible that our patient developed a local recurrence.

Radiographs may show an ill-defined soft tissue mass adjacent to but separate from bone.[3] Calcification is commonly seen. Although rare, periosteal reaction or cortical erosion can be found.[3,7] These radiological features are possibly suggestive of a malignant tumor involving the periosteum or cortex. On the other hand, in our case, although periosteal or cortical reaction was not seen, calcification developed to a size twice that seen initially within a period of 4 weeks. Because of its rapid growth, it was impossible to rule out extraskeletal osteosarcoma based on radiological features.

Histopathologically, the lesion is characterized by the following features: (1) localization in the subcutaneous tissue without muscular involvement; (2) a disorderly multinodular growth pattern with indistinct borders; (3) a fibroblastic proliferation showing variable degrees of cellular atypia; and (4) haphazardly arranged osseous trabeculae without the zoning phenomenon of myositis ossificans.[3] The present lesion exhibited most features commonly seen in previously reported cases.[1,3–5,8] In addition, an incomplete zoning phenomenon was observed in our case as well as in the cases reported by Sleater et al.[6] This incomplete development might occur because the constraints of the tissue planes of the digit do not allow for the full expansion and expression of the zoning phenomenon.

In conclusion, it is important for the clinician and pathologist to recognize the presence of fibro-osseous

pseudotumor of the digits and to treat accordingly. Aggressive surgical resection should be avoided.

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