Fibro-Osseous Pseudotumor of Toe: A Case Report

Ayak Parmağında Fibro-Osseöz Psödotümör

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ABSTRACT Fibro-osseous pseudotumor (FOPT) is a benign soft tissue tumor of the hand characterized by bone differentiation zones in proliferated fibroblasts. The etiology of FOPT is not yet known. In young adults, it occurs as a fast-growing, swollen and painful tumor particularly in hand's proximal phalanx. The tumor is thought to be related to myositis ossificans. In order to avoid unnecessary radical surgery and amputation, its differential diagnosis from extraskeletal osteosarcoma is critical. We report here the case of a 30-year-old female patient with post-traumatic FOPT. At the dorsum of the left fifth toe, a partially mobile grayish-purple 0.5 cm tumor with ulcerated surface was noted. In the histopathological examination, irregular tumoral tissue was observed in the ulcer base and random bone formation at the base of the lesion. The tumor consisted of slightly pleomorphic fibroblastic cells with indistinct cytoplasmic boundaries and large nuclei, some of them containing nucleoli. Osteoblasts without atypia were also present around the trabeculae. Although FOPT is reported to be a lesion in the hand, there is a case of FOPT in the foot was presented in the literature before. In this case report, we present the second case of FOPT in the foot in English literature.

Key Words: Granuloma, plasma cell; toes; osteosarcoma

ÖZET Fibro-osseöz psödotümör (FOPT) elde nadir görülen, prolifere fibroblastların içinde kemik farklılaşma alanlarının olduğu benign yumuşak doku tümörüdür. FOPT'ün etiyolojisi bilinmemektedir. Tümör genç yetişkinlerde özellikle el proksimal falanks seviyesinde hızlı büyüyen eritemli, şişkin ağrılı kitle şeklindedir. Tümörün myozitis ossifikans ile ilişkili olduğu düşünülmektedir. Gereksiz radikal cerrahiden ve amputasyondan kaçınmak için iskelet dışı osteosarkom ile ayırıcı tanısı son derece önemlidir. Sunduğumuz olgu 30 yaşında kadın hastadadır. Tümör travma sonrası gelişmiştir. Sol ayak beşinci parmak dorsal tarafında 0.5 cm büyüklükte, yüzeyi genel olarak ülsere gri-mor renkte kısmen hareketli kitle tespit edildi. Histopatolojik incelemede ülser zemininde düzensiz sınırlı tümöral doku görüldü. Tümörde sitoplazma sınırları tam olarak ayırt edilemeyen, bazıları nükleolus içeren, iri nükleuslu, hafif pleomorfik görünümlü fibroblastik hücrelerin olduğu zeminde rastgele dağılmış kemik oluşumları görüldü. Kemik trabekülleri çevresinde atipi içermeyen osteoblastlar da mevcut idi. Elde gelişen bir lezyon olarak bildirilmesine karşın, literatürde sunulmuş ayakta gelişen bir FOPT olgusu da vardır. Burada sunulan olgu, İngilizce literatürde günümüze dek sunulan ikinci FOPT olgusudur.

Anahtar Kelimeler: Granülom; ayak parmakları; osteosarkom

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Fibro-osseous pseudotumor (FOPT) is a rare benign soft tissue tumor of hands which is characterized by foci of osseous differentiation within the proliferated fibroblasts.[1-3] Myositis ossificans and FOPT are included in the same category in the World Health organization (WHO) new classification of soft tissue tumors.[2,4,6] FOPT is seen in young adults as an

erythematous, swollen, painful soft tissue tumor particularly in the proximal phalangeal level.[1,3] The differential diagnosis of this tumor from extraskeletal osteosarcoma is important.[1,3-5,7,8] In our study, the diagnosis and treatment of a FOPT in the 5th toe is evaluated and the importance of differential diagnosis is emphasized.

CASE REPORT

A 30-year-old female presented to our Orthopedics and Traumatology clinic in December 2005 with a 4-month history of a painful and slightly swollen bunion on her fifth left toe. She reported having bashed her foot into a rock while swimming one month before the bunion appeared. The physical examination revealed a 0.5 cm, partially mobile and painful tumor tied to the surrounding tissue at the dorsum of the middle phalanx of the left fifth toe. The skin on the tumor was grayish-purple in color. No local temperature elevation was present around the tumor. Periosteal thickening in the middle phalanx of the fifth toe and cortical destruction was not noted in the plain radiograph of the left foot. A slightly increased soft tissues shadow was noted at the dorsal aspect of the fifth toe. Calcification was not seen in the soft tissue. Blood count yielded normal white blood cells, erythrocyte sedimentation rate and C-reactive protein. After obtaining informed consent, the patient was operated for total excision of the tumor.

A 0.2 inch grayish-purple mass with distinct boundaries was observed macroscopically. There was 0.2 cm intact skin surrounding the mass. Following the fixation, the entire material was embedded in paraffin and five-micron-thick sections were stained with hematoxylin-eosin for examination. In the histopathological examination, a large ulcerated area was seen on the surface of the biopsy specimen. Tumoral tissue with multinodular growth pattern and indistinct boundaries was observed at the ulcer base. Haphazardly arranged osseous trabeculae were noted in the fibroblastic cell base in the tumor. Nuclear atypia or mitotic activity was not present in the fibroblastic cells and osteoblasts around the trabeculae (Figure 1). Inflammation was observed around the ulcerated area. Immunohistochemically, fibroblastic cells expressed vimentin (Vimentin, Clone SP20, NeoMarkers) (Figure 2). Smooth muscle actin (Actin, Smooth Muscle Ab-1, Clone 1A4, Neomarkers) or desmin (Monoclonal Mouse anti-Human Desmin, Clone D33, Dako) expression was not detected. Microscobically 0.2 cm intact skin tissue was observed around the lesion.

On 24th month control of the patient, no complaints or recurrence was present.

DISCUSSION

Fibro-osseous pseudotumor of the fingers, a benign lesion with heterotopic ossification, was first described and differentiated from other benign or ma-

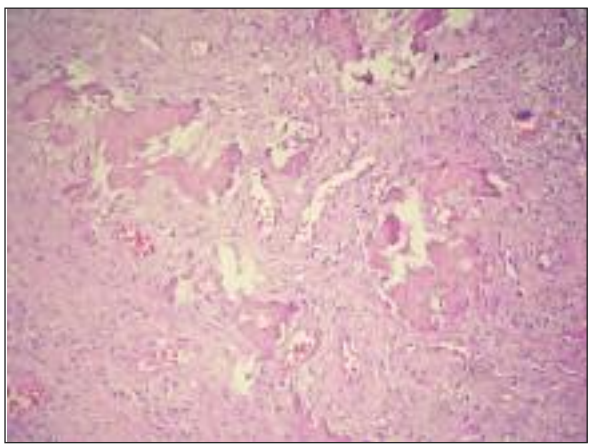

FIGURE 1: Spindle-shaped proliferated fibroblastic cells and osteoid formations (HEx200)

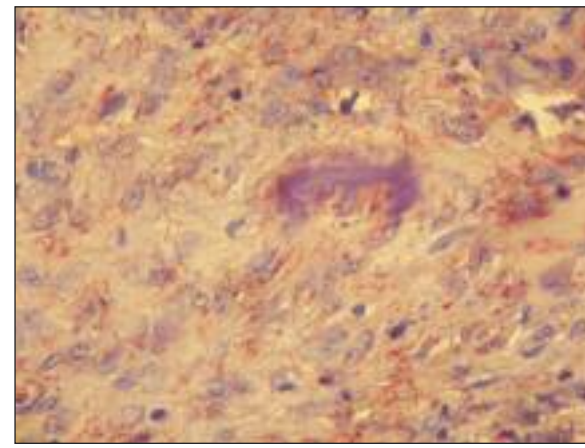

FIGURE 2: Vimentin expression in fibroblastic cells (AEC x 400).

lignant fibro-osseous lesions in 1986 by Dupree and Enzinger.[1] In previous studies, FOPT was reported as florid reactive periostitis of the tubular bones of the hand or foot, pseudomalignant osseous tumor in soft tissue, or parosteal fasciitis. The etiology of FOPT is not yet known; however, the tumor is commonly accepted to be a rare subcutaneous variant of myositis ossificans developing in the skeletal muscles.[1,2] Approximately 40% of the cases have a trauma history.[1,8] The tumor develops in several weeks or months, and its most common clinical symptoms are pain and bunions. The fingers may lose their function at times.[1,3,6]

The tumor is generally seen in the fingers.[1,3,4] Of the 21 subjects examined by Dupree and Enzinger and diagnosed as FOPT between 1949 and 1980, 20 had the tumor in their fingers and only one in the toes.[1] The tumor was identified in the proximal phalanx in six cases, in the distal phalanx in three cases, and in the middle phalanx in one case. Localization was not known in 11 subjects.[1] De Silva and Reid studied 14 cases of FOPT between 1962 and 2002; all of these tumors were located in the hands.[4] Nalbantoğlu et al. reported a fibro-osseous pseudotumor case in the wrist.[6] In our case, the patient had a trauma history, the painful tumoral tissue at the dorsal aspect of the fifth toe swelled slightly in the past 4 months.

The radiological appearance of the tumor varies by stage of development. Slightly more than half of the cases display increased density in the soft tissue caused by the tumor. In the remaining cases, soft tissue is calcified. Calcification is diffuse or irregular in a similar fashion to myositis ossificans.[9]There is no increase in peripheral density.[1] Periosteal thickening is seen in some cases, while cortical erosion is present in fewer cases.[1,3,4] In our case, the plain radiograph of the left foot illustrated a slight increase in soft tissue shadow at the dorsal aspect of the fifth toe. The bony structure was normal.

Histopathologically, fibro-osseous pseudotumor has four main morphological features: The tumor is in the subcutaneous region and has no relation to muscle and bone; it has irregular boundaries and displays a multinodular growth pattern; there is fibroblast proliferation and may include cellular atypia at differing levels; and randomly scattered osseous trabeculae and osteoid formations exist between these cells.[1,3,7] Despite irregular growth pattern and atypical fibroblastic cells, cellular atypia is not observed in the osteoblasts. No atypical mitotic figures are seen in fibroblasts or osteoblasts.[1]

The clinical and pathological differential diagnosis of FOPT should primarily and particulary include extraskeletal osteosarcoma.[1,3,5] On the other hand, differential diagnosis should also include bizarre parosteal osteochondromatous proliferation (Nora's lesion), osteochondroma and myositis ossificans.[1,2,4]

Due to aggressive growth, hypercellularity and cellular atypia, FOPT may be confused with extraskeletal osteosarcoma, thus making differential diagnosis critical.[1,3-5] The differential diagnosis of the two lesions may be particularly difficult in the early stages of the tumor. In a study conducted on 21 mature FOPT cases, Dupree and Enzinger reported that six had been initially misdiagnosed as either parosteal or extraskeletal osteosarcoma.[1] Two cases had been treated with finger amputation and one with large radical resection. De Silva and Reid similarly reported that one of the 11 FOPT cases they studied had been initially misdiagnosed as osteosarcoma, and finger amputation had been performed.[4] Clinically, extraskeletal osteosarcoma occurs rarely under the age of 35 and in hand fingers. It is mostly seen in the lower extremity with a high incidence in the thigh. Conversely, FOPT generally develops in young adults and is common in the fingers.[1,3] The most commonly affected finger is the index finger, followed by the 3rd and 4th fingers and the proximal phalanges. Histopathologically, extraskeletal osteosarcoma is characterized by more pleomorphic hyperchromatic cells and mitotic figures.[1,3,4]

Single or repeated episodes of blunt muscle trauma may lead to myositis ossificans. The lesion is frequently seen in adolescents and young adults, generally in the quadriceps, adductors, deltoid and brachialis muscles. The initial irregular radioden-

sity detected in the radiographs turn into bone formation as the lesion matures.[9,10] Microscopic examination reveals a characteristic zonal pattern. In other words, fibroblasts, myofibroblasts, vascular proliferation, chronic inflammation cells, and immature tissue resembling nodular fasciitis or granulation tissue containing macrophages exist in the center. At the periphery, there is an ossified area surrounded by osteoblasts and more mature tissue containing fibroblasts.[2,4]

Osteochondromas are the most common type of bone tumors. They generally develop in the metaphysis of long bones and occasionally in flat bones such as the pelvis and scapula. They project from a normal bone with a bony growth surrounded by a cartilage cap. A fibrous tissue capsule or bursa typically covers the cartilage surface. The diagnosis of osteochondroma can be made easily from plain radiographs. The medullar canal of normal bone continues at the base of the lesion.[10]

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 fingers and osteochondroma. Nora's lesion presents as well-delineated mass attached to the bone surface. Histologically, it has new bone formation surrounded by a cartilage cap at the periphery.[2,3]

Here we presented the case of a young adult. Clinically, the patient's age and the location of the tumor did not suggest extraskeletal osteosarcoma. Histopathologically, no cellular pleomorphism or atypical mitotic figures were seen that suggested osteosarcoma. Likewise, zonal pattern which is characteristic of myositis ossificans was not seen either. Our patient's lesion was not attached to the bone surface and no definite cartilage cap or chondrocytes were observed.

Total local excision is reported to be an excellent treatment for FOPT.[1-3] Recurrence may be seen at the end of 1 or 2 years following the initial excision.[1,4] Dupree and Enzinger identified recurrence one year after the initial excision in one of the seven cases.[1] The tumor was re-excised and did not recur in the following 8-year period. Again De Silva and Reid identified recurrence in one case 24 months after the initial excision.[4]

In the case reported here, the tumor was totally locally excised. No complaints or recurrence were present at 24 months follow up.

Fibro-osseous pseudotumor is a rare benign lesion which usually occurs in the fingers. Even though it is rare, this lesion should still be considered in painful tumoral formations that develop over weeks or several months in the toes. Its differential diagnosis from extraskeletal osteosarcoma is also important. The case we report is the second one in the English literature that reports a tumor in the toes.

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