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Case Report

Fibromatosis of the First Toe; Case Report

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Abstract

Extra-abdominal fibromatosis is a rare, aggressive monoclonal fibroblastic proliferation of musculoaponeurotic tissues. There are two types: superficial tumors, such as Dupuytren's disease, Lederhose's disease, or penile fibromatosis, and deep tumors, which are called aggressive desmoid tumors because of their local behavior. The term desmoid, derived from the Greek "desmos" meaning band, was initially applied by Müller because of its tendon-like consistency. They affect with a slight predominance the population between 25 and 35 years of age, representing about 0.03% of all neoplasms of the skin and parts blandas.se a 36-year-old female patient with a history of apparent health who reports an increase in volume of the thick artery of the left foot that causes pain and difficulty in wearing closed shoes for 13 years of evolution. It was decided to admit him for study and treatment.

Keywords: Desmoid Fibromatosis; Desmoid Tumor; Commonly Called Aggressive Fibromatosis

Introduction

Extra-abdominal fibromatosis is a rare, aggressive monoclonal fibroblastic proliferation of musculoaponeurotic tissues [1,2]. There are two types: superficial tumors, such as Dupuytren's disease, Lederhose's disease, or penile fibromatosis, and deep tumors, which are called aggressive desmoid tumors because of their local behavior [1]. The term desmoid, derived from the Greek "desmos" meaning band, was initially applied by Müller because of its tendon-like consistency. They affect with a slight predominance of the population between 25 and 35 years of age, representing about 0.03% of all skin and soft tissue neoplasms [1-3]. Desmoid tumors do not metastasize [1,2,4]. According to their anatomical location, they are classified as: extra-abdominal fibromatosis (60%); abdominal wall fibromatosis (25%) and intra-abdominal

fibromatosis (8-15%), the latter being the most biologically aggressive, due to its ability to infiltrate both pelvic and abdominal organs, [4].

Case Report

A 36-year-old female patient with a history of apparent health who reported an increase in the volume of the thick artery of the left foot that caused pain and difficulty in wearing closed shoes for 13 years of evolution, came to our clinic where it was decided to admit her for study and treatment.

Imaging study

A simple AP x-ray of the left foot, selective and magnified, showed irregular periostitis of the internal and posterior cortical of the distal phalanx of the 1st toe with increased volume and density of the adjacent soft tissues.

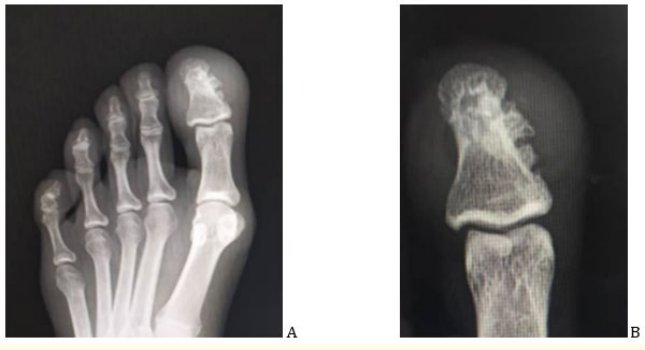

Figure 1: A and B: Periostitis of the internal cortical of the distal phalanx, 1st finger with soft tissue mass.

Simple CT scan both feet, MPR. There is evidence of soft tissue mass towards the internal and plantar region of the distal phalanx of the 1st toe, hyperdense nodular and polylobed with associated focal periostitis.

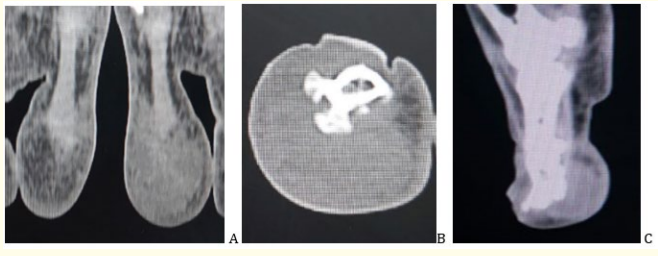

Figure 2: A-C The presence of fibroid of the 1st toe of the left foot with focal periostitis was observed.

Surgical procedure

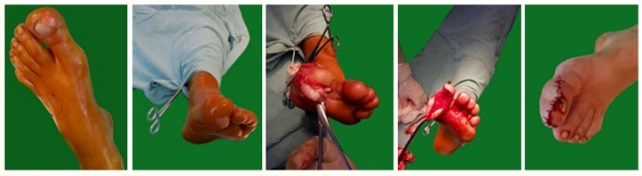

Figure a

Anatomical pathological findings

- **Macro:** Several masses of tissue, the largest of which is 4 x 3 x 2cm, irregular, whitish. To the cut of fibroelastic surfaces, swirling surfaces.
- **Micro:** Specimen formed by proliferations of mature fibroblasts that tend to form nodules and are grouped in fascicles surrounded by abundant collagen fibers.

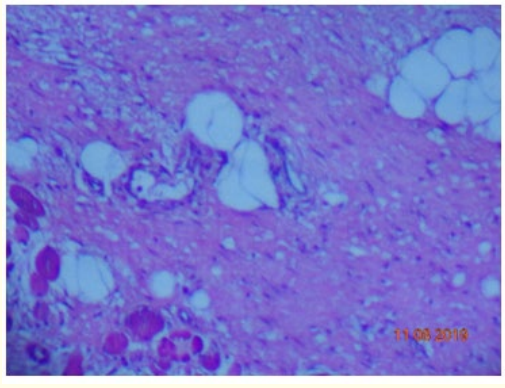

Figure 3: Panoramic view of mature fibroblasts arranged in fascicles. (4x).

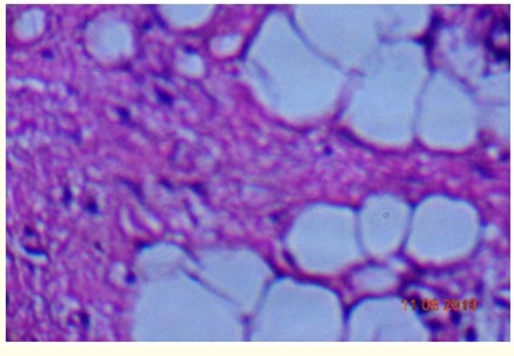

Figure 4: Mature fibroblasts arranged in fascicles surrounding adipocytes. (10x).

Discussion

Deep extraabdominal fibromatosis is called an aggressive tumor because of its local behavior [1]. They affect the population between 25 and 35 years of age with a slight predominance, although, in many series, no differences are observed according to age; they represent about 0.03% of all skin and soft tissue neoplasms, [1-3]. do not metastasize, [1,2,4]. however, they are classified as neoplasms of intermediate malignancy, [5-7]. Due to its high ability to infiltrate adjacent structures and organs and the high local recurrence rate of 20%-64% after surgical resection,6,7 and repeat surgery often entails substantial morbidity. Even after a complete resection with neoplasm-free margins, this is why it is also known as deep fibromatosis or aggressive fibromatosis [3].

Non-surgical modalities such as chemotherapy, radiation therapy, and hormone therapy have been considered an alternative treatment option.(7) On the other hand, some authors advocate a more conservative approach, which is known as the "wait and see" strategy, as an initial approach [7,8]. These tumors arising from connective tissue, muscle, fascia, or aponeuroses are rare, with a reported incidence of approximately 2-4 per million per year, with 3% of FD cases among all soft tissue tumors [2,8].

FD Occurs more frequently in women than in men [1,2,9,10].

Between puberty and age 40, those under the age of 30 have a higher recurrence rate [11].

Imaging techniques allow for a differential diagnosis of other tumors that affect soft tissues. X-ray can identify whether or not the tumour involves bone structures, MRI can evaluate soft tissue tumours, CT scan can assess retroperitoneal sarcomas, and the definitive diagnosis is with biopsy and histopathological study [2].

When there is a palpable mass, the first imaging test requested is ultrasound. Their findings are non-specific, although they usually present as a predominantly hypoechogenic mass with subsequent acoustic reinforcement in 75% of cases. Due to their high hypoechogenicity, they can sometimes be misinterpreted as cystic lesions, for which color Doppler can be useful, although those with fibrous prodominance sometimes do not show flow [10].

CT scans are visualized as isodense lesions to the muscle with some areas of higher density due to the existence of increased collagen deposition and others of lower density of myxoid content. The contrast is variable and can sometimes be intense. In fibromatosis of the thorax and abdomen, CT may be more useful due to the ventilation artifacts that MRI can present, which does not occur in other locations such as the limbs [10].

MRI is the gold standard test for diagnosis and follow-up. Due to its high tissue resolution, it allows the delimitation of the margins for an intervention, as well as the adequate establishment of their anatomical relationships [10]. The imaging findings will allow us to suggest its diagnosis and propose the differential diagnosis with other tumors such as undifferentiated pleomorphic sarcoma (main differential diagnosis), fibrosarcoma, lymphoma, and densely calcified masses.

In the case of our patient, due to the age, location and radiological appearance of the lesion, the differential diagnosis was made with the following most frequent bone and soft tissue lesions in the phalanges of the hands and feet.

- **Osteochondroma**: It is a bony outgrowth with a covering of hyaline cartilage that originates on the surface of the bone. They can be single or multiple (multiple hereditary exostoses). Radiologically, they present as a bony protrusion protruding from the surface of the bone, containing cancellous, cortical bone in continuity with the underlying bone. Clinically, they usually present as a slow-growing, asymptomatic mass [12].
- **Periosteal chondroma**: A tumor composed of hyaline cartilage that originates adjacent to the cortical surface, below the periosteum. On radiography, it presents as a mass of soft tissues with some erosion of the adjacent cortical. Thickening of the cortical at the proximal and distal margins of the tumor is typical, which in turn may produce some periostitis. Calcifications are present in about half of cases [12].
- **Subungual exostosis** is a benign tumor that originates in the distal phalanx, below or adjacent to the nail bed. Its radiological appearance is similar to that of osteochondroma, although it is distinguished histologically by being formed by trabecular bone covered with fibrocartilage. It may be asymptomatic or present with pain, swelling, and ulceration of the nail bed or surrounding soft tissue [12].

- **Glomus tumor**: A benign tumor that originates as a result of hyperplasia of the neuromyoarterial glomus body usually present in the region of the nail bed. They are typically extraosseous that can secondarily affect the bone, although they can be intraosseous. On radiography, soft tissue glomus tumors may present with well-defined erosion in the underlying bone (sometimes with a sclerotic margin), predominantly in the distal phalanx. Clinically, they present with pain, which when triggered after a slight trauma or exposure to cold is quite suggestive of a glomus tumor [12].
- **Gouty topho:** It is caused by the deposition of monosodium urate crystals. In the phalanges, this deposit ("topho") occurs in the soft tissues and results in erosions in the adjacent bone. It does not affect the joint space [11].

Fibromatosis should be differentiated in pathological anatomy from aponeurotic calcifying fibroma, desmoid-type fibromatosis, cellular fibrous histiocytoma, monophasic fibrous synovial sarcoma, and fibrosarcoma. Aponeurotic calcifying fibroma occurs in both palms and soles, but shows infiltrating growth that differentiates it from superficial fibromatoses that occur in that topography. Desmoid fibromatosis is also infiltrating and, although the location in the trunk is the most common, 2% of cases can occur in plants, as in our case. Sarcomas are usually deeper tumors and have features of malignancy [13].

In the case of our patient, it was decided to perform surgical treatment due to the presence of pain and difficulty in wearing closed shoes, continuous follow-up by outpatient clinic with favorable evolution after three months, there is no evidence of recurrence.

Conclusion

Desmoid-type fibromatosis (FD) is a rare tumor, considered benign but locally infiltrating, and can occur almost anywhere in the body. Despite not being metastatic, this neoplasm has a local recurrence rate of 20%-64% after surgical resection. There is no consensus on the optimal treatment of desmoid tumors, so there is no protocol for optimal treatment management, which remains a challenge.

Conflict of Interest

The authors of this paper certify that there are no conflicts of interest in the realization and review of this topic.

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