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Case Report

Extra-Abdominal Fibromatosis -Case Report

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Abstract

Desmoid-type fibromatosis, or desmoid tumor, is a locally aggressive, growing infiltrating myofibroblastic lesion with unpredictable clinical behavior. It was first described by MacFarlane in 1832 and named the desmoid tumor by Mueller in 1838. Aggressive treatment may be necessary when clinical symptoms such as localized pain are severe or affect appearance, with between 20% and 64% of patients experiencing recurrence. We present a 42-year-old female patient with a history of apparent health who reported an increase in volume in the posterior area of the left thigh of 3 months of evolution, causing numbness of the leg, pain and difficulty of mobility of the left knee and histopathologically it turned out to be an extra-abdominal desmoid tumor. The patient continues to be followed up by outpatient clinic.

Keywords: Fibromatosis, Histopathologically

Introduction

Desmoid-type fibromatosis, or desmoid tumor, is a locally aggressive, growing infiltrating myofibroblastic lesion with unpredictable clinical behavior. It can originate anywhere on the body, with extremities, abdominal wall and mesentery being the most common Sites. The peak of incidence is in the third decade. Desmoid tumors arise sporadically in about 90% of cases. The remaining 10% is family-owned [1]. It is a benign tumour condition, classically treated by a more or less radical and sometimes mutilating excision [2]. It is a borderline tumor with local invasion but rarely metastasizes at a distance [3,4]. It has a strong tendency to local invasion and local recurrence. The lesions are often sensitive and can limit the patient's mobility, especially if they are near a joint. The tumor can invade or compress local nerves, causing neurological symptoms such as numbness, pain, and loss of mobility. Aggressive treatment may be necessary when clinical symptoms such as localized pain are severe or affect appearance, although surgical resection has long been the first choice. [4] Between 20% and 64% of patients experience recurrence [4,5].

Case

A 42-year-old female patient with a history of apparent health who reported an increase in volume in the posterior area of the left thigh for 3 months, causing numbness of the leg, pain and difficulty in mobility of the left knee, came to our clinic where it was decided to admit her for study and treatment.

Imaging Result

Radiography

Increased volume of the soft tissues in the area of the shaft of the femur without bone alterations

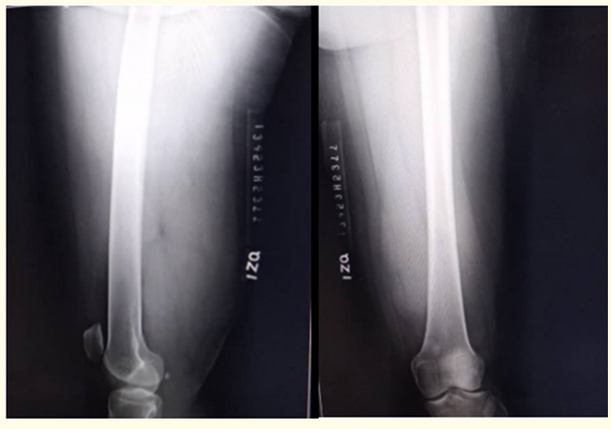

Figure a

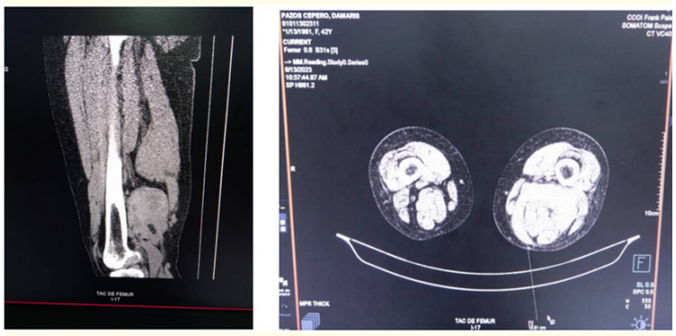

Figure b

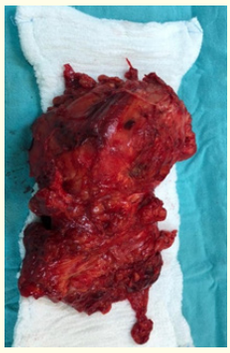

Figure d

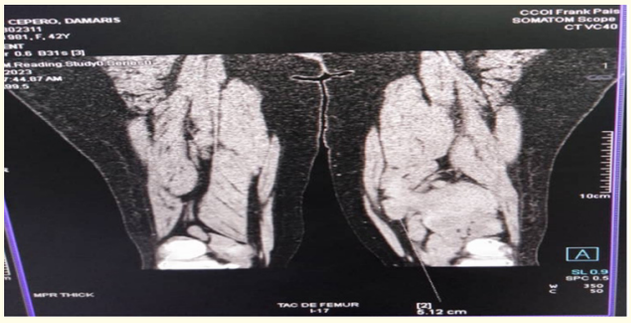

Figure c

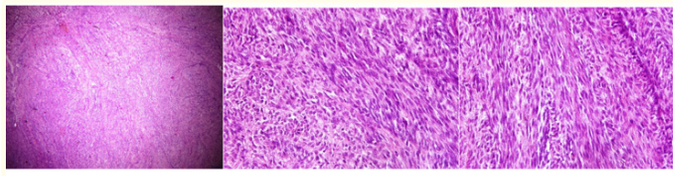

Figure e: Panoramic view of Desmoid Tumor (4x) Arrangement of cells in fascicles (10x) Clustered spindle cells arranged in fascicles (40x).

CT scan of thighs

The Somaton Scope team study is conducted.

At the level of the distal portion of the biceps femoris muscle in its long portion, a hyperdense mass of irregular shorts is observed, which extends towards the inner aspect of the thigh to the medial area relating to the semimembranosus muscle displacing the vascular bundle in its proximal part and encompassing it in the popliteal area, the mass displaces the sartorius muscle with a density ranging from 46 to 66 UH. In its extension towards the inner thigh, it occupies a fairly superficial path of approximately 10.5 cm in length x 5.1 cm in axial section.

No bone alterations are observed

ID. Rule out rhabdomyosacroma of the left thigh.

Anatomical pathological findings

Macro

In Pathological Anatomy, a fragment of irregular, grayish tissue measuring 12 x 7 x 7 cm is received. When cut, firm consistency, whitish, multinodular, swirling, well-defined surface

Micro

The histological study showed proliferation of spindle cells arranged in fascicles with densely populated areas, others less cellular, mixed with collagenous fibers. No presence of mitotic figures.

Immunohistochemistry

In the IHC study performed on our specimen, strong positivity for Vimentin and weak positivity for Desmin was observed. It is negative for Actin, Keratin, EMA, CD34 and CD117.

Anatomopathological Diagnosis: Desmoid Tumor

Discussion

Desmoid-type fibromatosis (FD) is a rare mesenchymal tumor, first described by MacFarlane in 1832 and named desmoid tumor by Mueller in 1838 [5,6]. This neoplasm is considered benign but locally infiltrating and can occur almost anywhere in the body. It happens sporadically [6]. Despite not being metastatic, this neoplasm has a local recurrence rate of 20%-64% after surgical resection, and repeat surgery often results in substantial morbidity [4,6]. Non-surgical modalities such as chemotherapy, radiotherapy and hormone therapy have been considered an alternative treatment option [6]. On the other hand, some authors advocate a more

conservative approach, known as the "wait and see" strategy, as an initial approach [6,7]. These tumors arising from connective tissue, in muscle, fascia or aponeurosis are rare, with a reported incidence of approximately 2-4 per million per year, among all soft tissue tumors there are 3% of FD cases [7-10].

FD Occurs more frequently in women than in men [9-11].

Between puberty and age 40, those under the age of 30 have a higher recurrence rate [12,13].

There is no consensus on the best treatment options [6,14]. Surgical excision with wide margins.

It is still considered the gold standard, but a recent paradigm shift towards more conservative ones due to the high recurrence rate (up to 60%) and associated morbidity. The 2019 global consensus guidelines of the Desmoid Tumor Working Group state that surgery should be avoided whenever possible and propose active surveillance as the first line [14].

There is a prognostic difference between intra- and extra-abdominal, and it is known that extra-abdominal development is rarely life-threatening [15].

Both abdominal and extra-abdominal localization can cause pain, and functional impairment as a result of joint contracture and compression of major neurovascular systems in the extremities. Although it does not metastasize, it can still be fatal if it compresses adjacent vital structures [16].

Treatment modalities include "watchful waiting," nonsteroidal drugs, anti-inflammatories, antiestrogens, chemotherapy, Targeted therapy, radiation therapy, surgical excision, and combination of the above. However, management remains a challenge [16].

Histologically, it resembles a low-grade fibrosarcoma and has a low mitotic index, so it can be said that its differentiation from sarcomas is of great importance. Compared to fibrosarcomas, aggressive fibromatosis presents with poorly defined borders, there is no presence of severe cellular atypia or mitosis, while low-grade fibrosarcomas have significant cellular atypia and abundant mitotic figures [17]. Keloids differ in that they have more mature hyaline collagen with edematized fibroblasts and tablets. Fibromatosis always has a higher degree of cellularity and more interstitial collagen than a myxoma. Fasceitis has a chronic, scattered inflammatory infiltrate and is a small lesion of variable cellularity [18]. Other entities must be differentiated from fibromatoses such as low-grade fibromyxoid sarcoma, myofribosarcoma, solitary bone tumor, myofibroma, and nodular fasciitis [19].

In the case of our patient, it was decided to perform surgical treatment due to the presence of pain and neurovascular manifestations due to compression of the sciatic nerve and the vascular bundle at the level of the popliteal area, continuous follow-up by outpatient clinic with favorable evolution after three months, there is no evidence of recurrence and the neurological symptoms continue with the patient's perception of improvement and she does not present motor deficit.

Conclusion

Desmoid-type fibromatosis (FD) is a rare mesenchymal tumor, is 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 extra-abdominal desmoid tumors (EDTs), so there is no protocol for optimal treatment management, which remains a challenge.

Conflict of Interest

None to declare.

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