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Desmoid tumor of the foot: a case report and literature review

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ABSTRACT

A desmoid tumor (DT) was first described by MacFarlane in 1832. DTs are rare tumors, corresponding to only 0.03% of all neoplasia and less than 3% of all soft tissue tumors. Most of the tumors occur in the abdomen and presentation in the extremities is uncommon. Here, we present a review of the literature and the 27th case of DT of the foot.

Keywords: aggressive fibromatosis; foot; orthopedics

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Introduction

MacFarlane first described a desmoid tumor [DT] in 1832 [1]. DT is also known as fibromatosis or desmoid-type fibromatosis [2]. Cases of DT are rare and in spite of its benign nature, local recurrences are common. It is known to invade nearby structures; however, it rarely infiltrates the bones [3,4]. The origin of the DT has not been completely elucidated, but prior trauma and endocrine and genetic components have been identified as predisposing factors [5]. DT can affect approximately 4 million people per year, mostly women between the third and fourth decade of life, indicating that there is a hormonal influence on the tumor [3]. When DT manifests in extra-abdominal regions, the main sites of involvement are the limbs and waist, with involvement of the extremities being extremely rare [4.6].

The development of DT is unknown due to lack of common pattern. Some tumors have accelerated growth, others do not change, and some regress without any treatment after its diagnosis. This lack of parameters, due to the heterogeneous manifestation of the tumor, makes the treatment difficult [4]. The management of this type of tumor is usually by surgical intervention. However, despite surgery being the most common treatment, local recurrence may occur with an 80% chance after the removal of the primary tumors [3,6]. In addition to other adjuvant treatments, there is a period called active surveillance, which is intended to observe the behavior of the tumor for 1 to 2 years with the aid of nuclear magnetic resonance [NMR] as spontaneous regression may occur in cases of extra-abdominal DT, which could modify the treatment of the patient [3,7].

Literature Review

We performed a search of the PubMed database using the descriptor “aggressive fibromatosis,” on June 23, 2019 without a time limit, seeking to identify all published cases of DT on the foot. The search resulted in 2039 articles, of which 1613 were excluded after evaluation of the title and another 321 were excluded after evaluation of the abstract. Furthermore, after reading the full text of the 105 remaining articles, we selected 17 studies, involving a total of 26 patients with DT on the foot, for this review.

The first description of desmoid tumor of the foot was made in 1942 by Pearman and Mayo, who presented a clinical and pathologic analysis of 77 cases, of which only 1 case pertained to DT on the foot [1]. Mehrotra *et al.* [2000] evaluated 36 cases of fibromatoses in the extremities in their study, of which 2 cases pertained to feet involvement. All cases were treated by surgical excision and without adjuvant therapy. They reached the conclusion that the control of fibromatoses in extremities is still complex. Despite the evaluation of clinical and histological aspects, it was not possible to determine or predict the risk of local recurrence of the fibromas after excision [15].

Karakousis *et al*. [1993] followed a series of 26 patients with DT from July 1977 to May 1991, of which 3 patients had tumors in the plantar fascia of the foot. The results of the study suggested that DT of the trunk and the extremities are highly curable especially if a combination of treatment modalities is adopted during the monitoring [16]. Dalén *et al*. [2003] performed a retrospective study on 30 patients with DT and performed patient follow-up for 20 years after treatment. Of these 30 patients, only 1 patient has a fibroma on the foot with multifocal presentation [foot/leg/pelvis] [10].

McDougall and McGarrity [1979] reported 3 cases of extra-abdominal DT. One cases pertained a 25-year-old woman who initially developed a tumor in the right foot in the region of the forefoot, then in the abdomen, and later in the right thigh with multiple recurrences after surgical excision. After 19 years of follow-up the patient had no more relapses, but some sequelae to the several surgeries performed. MacGill *et al*. 2011 described a case report of a 34-year-old woman who presented a desmoid

fibroma on the foot, treated by wide surgical excision without complications and [9] without recurrence after 28 months [11].

Henrik A. Schrøder & Siems J. Siemssen [1985] report a case of DT in the extremities of a father and son, but only the father's foot was affected. From the age of 10 years, the father had recurrent ulcerations in his right foot after a trauma and had undergone 2 surgeries on the foot. At the age of 60 years, a mass began to grow, which increased rapidly in size over 5 months, reaching 10 × 15 cm at the time of diagnosis. Due to the extension of the tumor, the involvement of bones and nerves, transtibial amputation was performed and no recurrence occurred in the 3 years following the follow-up [9].

Ronald Barbella and Ira M. Fox [1996] published a case of a 22-year-old woman who presented with 1-year history of pain and a large mass in the left foot plantar region. The patient reported having already removed another mass, however in the dorsal region of that same foot, 4 years before the reported episode. The treatment option chosen by the patient was the surgical excision of the tumor, due to the tumor causing pain and limiting her movements [5].

Brenner and Rammelt [2002] reported 2 cases of DT; 1 case pertained to a 37-year-old female patient with multifocal DT in the abdomen and the other case pertained to a 67-year-old woman with DT on the left foot. Both the cases were treated with extensive surgical resection and reconstruction with tissue transfer [18].

The case presented by Stengel *et al.* [2008] was that of a 31-year-old woman with DT on the left foot evolving since childhood. Multiple surgeries, recurrences and disabling pain marked this case, which was finally treated with pegylated alpha interferon for 51 months. This resulted in tumor stabilization and pain reduction from the second application and the patient returned to daily work activities [19]. Kolih *et al.* [2012] reported a case of a 19-year-old male patient with a history of increased volume in the posterior face of the left thigh 2 years prior to the study. Excisional biopsy was performed with a diagnosis of DT. However, after 60 days, 2 lesions were identified on the left foot, histologically compatible with recurrence of DT [14].

Prodinger PM *et al.* [2013] published a report with 27 cases of DT in the extremities, of which only 1 case pertained to a 11-year-old female patient with DT on the foot who had 2 recurrences during the 126-month follow-up [8]. Subsequently, Kolur Puttaswamy Raju *et al.* reported a case of a 20-year-old male patient who presented a small mass on the left foot, which grew over 2 years, extending from the sole to the dorsal aspect of the foot, hindering his walking. The treatment was surgical, with amputation of the second and third toe, but after 2 years he had no recurrence and began to walk normally [4].

A study reported by Yoon G. *et al.* [2014] reviewed 11 patients, treated between 2005 and 2011 with oral methotrexate and chemotherapy. Two cases occurred on the foot and have been followed for at least 12 months. No deaths occurred during the follow-up period, but the recurrence rate was 33% [12]. Another group of Indian researchers published a case report in 2015 of a young 30-year-old woman with a history of swelling in her right foot that had evolved 10 years ago. After investigation, she was diagnosed with primary foot injury and pulmonary metastasis, and chemotherapy was chosen, however, the patient died 5 months after this diagnosis [13].

Wirth L., *et al.* [2018] reported that from 1981 to 2014, their institution treated 44 cases of DT in the extremities and trunk, 6 of which were located in the foot. Various treatments were performed on these 44 patients, depending on each case, including surgery, radiotherapy, therapy with NSAIDS and chemotherapy, but the treatment for the foot DT was not specified [6]. In the same year, Qinqin Liu, MDa *et al*. presented a case of a 41-year-old female patient who was hospitalized with a painful mass on the right foot with continuous growth for 3 years. The patient was treated for the DT with incomplete resection,

continuing with monitoring for 16 months without local recurrence [3].

Case Presentation

This case pertains to a 32-year-old male patient with a slow-growing tumor lesion on the dorsum of the left foot for 3 years, evolving with pain and difficulty in wearing footwear [Figures 1 and 2]. X-ray revealed a volume increase of soft parts, without changes to the bone. NMR showed voluminous expansive lesion on the dorsum of foot with lobulated aspect measuring 8.1 × 7.5 × 3.5 cm and showing hypersignal during T2-weighted imaging [Figures 3]. A biopsy led to the diagnosis of desmoid tumor/aggressive fibromatosis. Surgical treatment was suggested; it was performed on August 31, 2016, with complete excision of the lesion, but without pathological free margins. During the postoperative follow-up, after about a year, recurrence of pain was observed and a volume increase. New magnetic resonance imaging [MRI] revealed a lesion with the same characteristics previously observed but with lesser extent, measuring 3.4 × 2.6 × 1.2 [Figures 4].

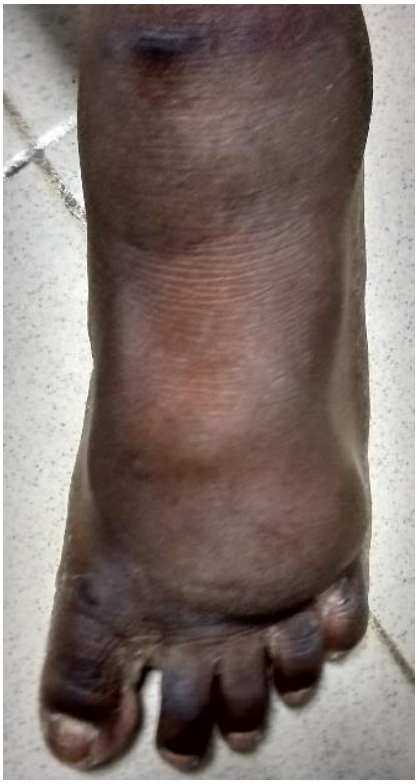

Fig. 1. Pre-operatory antero posterior view of the foot

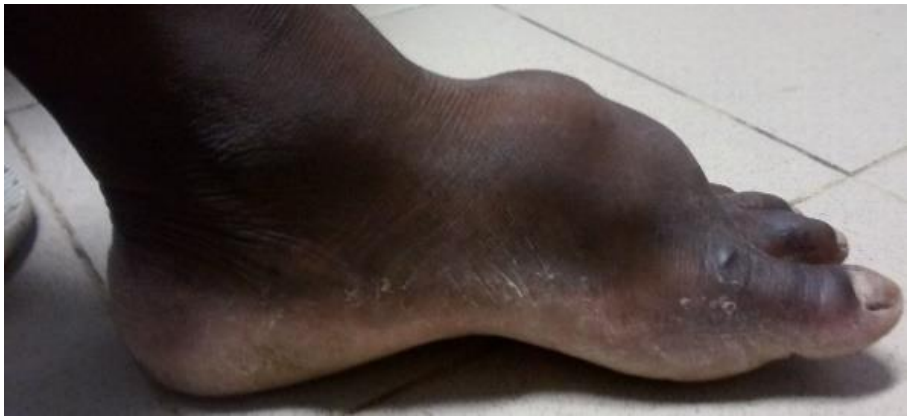

Fig. 2. Pre-operatory lateral view aspect of the foot

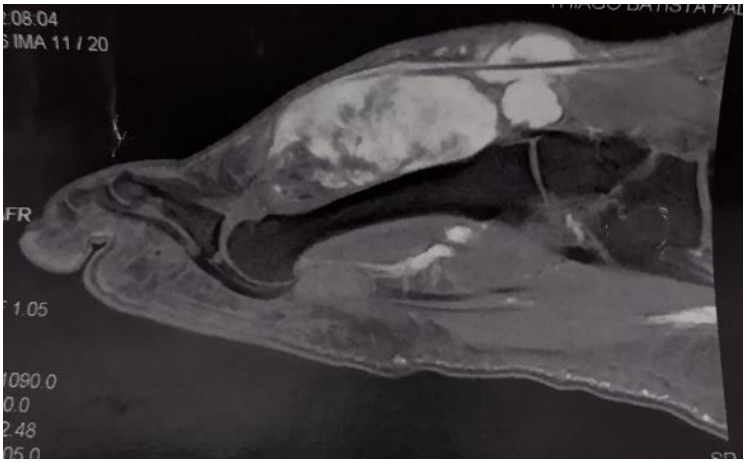

Fig. 3. MRI of the- sagittal T2 sequence

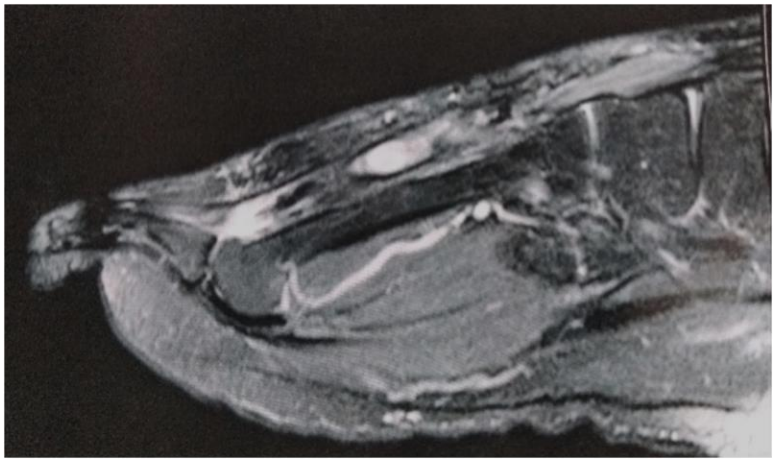

Fig. 4. 01 year pos-operatory MRI of the foot

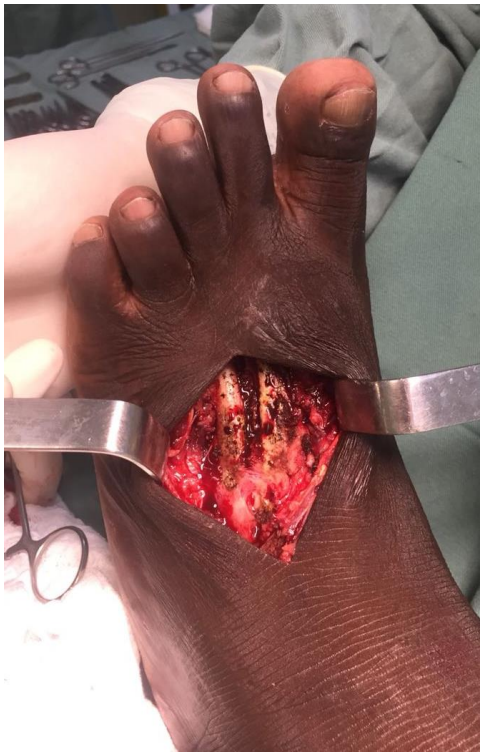

Fig. 5 Intra operatory view of the foot

A new surgical approach was performed, with complete excision of the lesion. The tumor was dorsally located on 2 occasions as the recurrence, as shown by preoperative MRI, having been used the dorsal longitudinal access on the dorsum of the forefoot and midfoot, on 2 occasions [Figure 5]. Currently, the patient is under follow-up without recurrence after 24 months, indicating an improvement of the pain.

The patient gave authorization for the publication of this article.

Discussion

DT on the foot is rare, having been reported only in 26 cases prior to the case presented here. The location of involvement of DT is a determining factor in the increase of local recurrence, being this recurrence extremely high for tumors of the extremities. The poor prognosis of DT in the extremities is also reported in the literature, having a worse outcome when compared to DT in other parts of the body [8]. This type of tumor has a large tendency to infiltrate in the adjacent structures, be they muscles, adipose tissue and even nerves or vessels [9].

The clinical presentation is a large mass, painless to palpation, well adhered, with little defined margins. Symptoms such as pain and paresthesia reveal that there is a commitment of the structures around the tumor, such as nerves. The fact that the tumor has an average size of 5 to 10 cm in diameter, may also hinder daily activities, as in the case of our patient who could not wear shoes [5].

MRI is the examination of choice for the diagnosis and characterization of tumors of the soft parts. In case of DT, MRI is also important for the clinical follow-up, along with computed tomography and ultrasound. MRI can aid in differentiating between DT and malignant lesions, such as weighing in T1 and T2. The non-fibrous lesion of the DT appears with less intensity than T2-weighted fat compared to a malignant lesion [10,11].

The key treatment for DT is local surgical excision with wide and safe margins, however, knowing that DT has an infiltrative pattern, most of the time a complete excision is technically difficult to obtain and tumor recurrence is frequent [12-14]. In these cases, and in those with bulky lesions, a viable solution is amputation [13]. There are non-invasive therapeutic options with limited results. Combined chemotherapy, radiotherapy, use of hormones and non-steroid anti-inflammatory drugs are therapeutic measures that have been used to reduce the tumor volume and the symptoms of the patients. More recently methotrexate in combination with chemotherapy presented positive results in the control of the DT. Wiess and lackman [1995] obtained an average of 77% of total or partial remission with the use of chemotherapy combined with methotrexate. However, in the patients studied by Van der Hul *et al.*, only 20% of the lesions responded to methotrexate.[11, 13-14]

Our patient is male, but most selected articles indicate that DT occurrence is more common in women. As in the studies discussed above, despite the complete excision of the primary tumor, recurrence of DT occurred after 1 year and we opted again for surgical treatment, which is in accordance with most of the data we found. There is no consensus in the literature on the management of TD in the feet. Due to its rarity, it is not possible to carry out large prospective studies comparing different techniques.

Conclusion

The literature review identified only 26 reported cases of DT located on the foot. The present report is the 27th case of DT on the foot and was treated surgically having evolved with locoregional recurrence. After the second surgical approach, the patient is in outpatient follow-up with improvement of in pain, function, and no signs of new tumor lesion.

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