

# Superficial acral fibromyxoma in a female patient: a case report

*Fibromixoma acral superficial em paciente do sexo feminino: um relato de caso*

## ABSTRACT

*The authors describe the case of a 75-year-old female patient, bearing a single asymptomatic, nodular lesion of fibroelastic consistency for three years, located on the left hallux. Following surgical excision of the lesion, the pathological examination and immunohistochemistry revealed a superficial acral fibromyxoma. This type of lesion was first described in 2001 and today there are approximately 100 cases in the literature. It is a benign mesenchymal tumor of slow growth, with a predilection for unguinal and peri-ungual regions. There are no reports of malignant transformation and recurrence has been associated with incomplete resection.*

**Keywords:** fibroma; neoplasm; nail diseases.

## RESUMO

Descreve-se caso de paciente do sexo feminino de 75 anos, apresentando há três anos lesão única, assintomática, nodular, de consistência fibroelástica, localizada no primeiro pododáctilo esquerdo. Indicada a exérese cirúrgica da lesão, o exame anatomopatológico e a imuno-histoquímica revelaram fibromixoma acral superficial. Esse tipo de lesão foi descrito pela primeira vez em 2001, e hoje há aproximadamente 100 casos na literatura. Trata-se de tumor mesenquimal benigno, de lento crescimento, com predileção por regiões ungueais e periungueais. Não há relatos de transformação maligna, e a recorrência tem sido associada à ressecção incompleta.

**Palavras-chave:** fibroma; neoplasias; doenças da unha.

## INTRODUCTION

Superficial acral fibromyxoma (SAF) was first described in 2001 by Fetsch et al. in a series of 37 cases.<sup>1</sup> Since then approximately 100 cases have been reported in the literature.<sup>2</sup> Yet this neoplasia is still poorly recognized by pathologists and dermatopathologists, partly due to its relatively uncommon occurrence, and partly because it has only recently been described.<sup>3</sup> It is a benign, slow-growing mesenchymal tumor, with no reports of malignant transformation or metastasis. The SAF has a clear tendency to involve the nail and periungual regions of the hands and feet. Middle-aged men are more frequently affected, with previously published case series suggesting that toes are more affected than fingers.<sup>4</sup> The usual treatment is complete surgical resection of the lesion, with a few cases of recurrence having been associated with incomplete resection.<sup>2</sup>

## Case Reports

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## CASE REPORT

A 75-year-old mulatto female patient sought care complaining of a “callus” in the foot for three years. She denied pain or trauma preceding the onset of the lesion. The physical examination showed nodules of approximately 1 cm, with a color similar to that of the skin, fibroelastic consistency, and slight scaling located in the medial nail fold of the left big toe. It also presented with constitutional melanonychia and desquamation around the nail of that toe (Figures 1 and 2). Dermoscopy showed intense subungual hyperkeratosis without the presence of specific structures and no vascularization (Figures 3 and 4). The patient underwent an excisional biopsy with the histology revealing a dermal mesenchymal lesion composed of stellate or spindle-shaped fibroblasts without atypia, permeated by capillaries in the mid the myxoid matrix. The immunohistochemical analysis revealed diffuse expression of CD34, focal expression of CD99 and EMA with an absence of expression of the S-100 protein, or desmin and actin of the smooth muscle tissue. The clinical pathological features associated with the immunohistochemical

profile defined the diagnosis of superficial acral fibromyxoma (Figures 5 and 6). The patient is still undergoing follow-up without signs of recurrence.

## DISCUSSION

Superficial acral fibromyxoma affects more men than women (2:1), typically between the ages of 14- and 75-years-old, with a mean age at diagnosis of 43 years.<sup>2,5</sup> The SAF tends to present as a slow growing, firm mass or nodule, almost always located in the toes and fingers.<sup>3</sup> However, it can less commonly affect palms, heels, ankles, and thighs.<sup>4</sup> The nail is involved in 50% of cases, with hyperkeratosis or onychomycosis. A history of trauma preceding the appearance of the lesion is rare.<sup>6</sup> The SAF is characterized by being painless, a fact that explains the delay of patients in seeking medical care.<sup>1</sup> It is histologically characterized by a well-circumscribed dermal or subcutaneous tumor with increased vascularization, constituted by fusiform or stellate cells embedded in alternating areas of fibrous and myx-



FIGURE 1: Skin-color nodular lesion of approximately 1 cm emerging from the medial nail fold of the left big toe

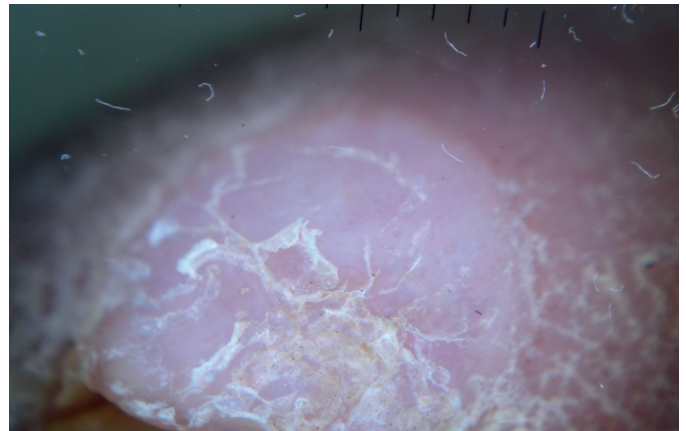


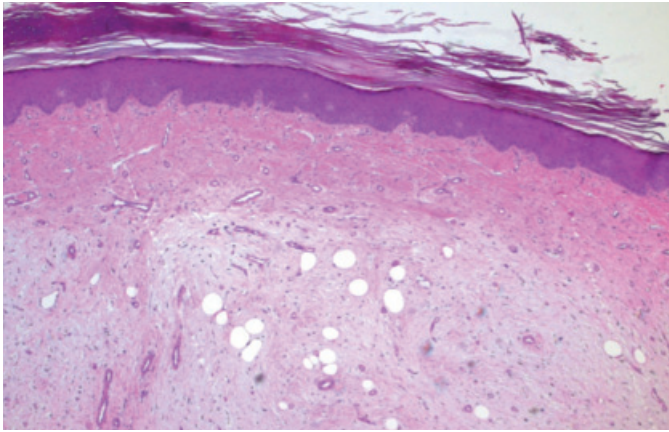
FIGURE 3: No structures or increased vascularity are seen under dermoscopy



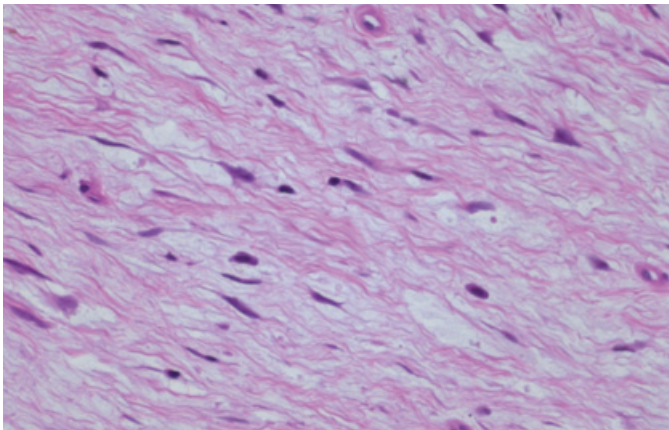
FIGURE 2: Detail of the skin-color nodular lesion, with fibroelastic consistency and presence of desquamation on and around it



FIGURE 4: Intense subungual hyperkeratosis is seen under dermoscopy



**FIGURE 5:** Circumscribed and well demarcated neoplastic proliferation located in the dermis (HE stain, 5x original magnification)



**FIGURE 6:** Neoplastic proliferation is composed of spindle cells without atypia, a mid myxoid stroma (HE stain, 40x original magnification)

oid stroma. The presence of significant nuclear atypia is rare and although it has been described in isolated cases, it causes concerns about the biological potential of the tumor. None of the tumors examined showed frank sarcomatous change, and no case of malignant transformation has been described in the literature.<sup>2,3</sup> The SAF is immunopositive for CD34, CD 99, and EMA, and negative for cytokeratin, melanocytic markers, SMA, and desmin.<sup>3</sup> The differential diagnosis of SAF should include consideration of ungual/periungual fibroma, acquired digital fibrokeratoma, low-grade fibromyxoid sarcoma, dermatofibroma, superficial angiomyxoma and myxoid neurofibroma.

The treatment of choice for SAF is surgical resection with free margins. Periodic monitoring is recommended after excision, and recurrence rates are estimated in the range of 10–24%.<sup>6</sup> Recurrence has been associated with incomplete resection.<sup>2,5</sup> The patient described showed no signs of recurrence of the lesion during a long follow-up. Although rare, SAF should be included in the differential diagnosis of tumors involving fingers and toes.<sup>5</sup> ●

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