

## Giant cell tumor of the tendon sheath: a rare periungual location simulating myxoid cyst\*

Renan Minotto<sup>1</sup>  
Aline Barcellos Grill<sup>1</sup>

Camila Britto Rodrigues<sup>1</sup>  
Roque Furian<sup>2</sup>

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**Abstract:** Giant cell tumor of the tendon sheath is a benign soft tissue tumor most frequent between the third and fifth decades of life. It can mimic and make differential diagnoses with several hand tumors. Definitive diagnosis and the treatment of choice are reached with complete resection and histopathological examination. Here we describe a case with clinical presentation similar to that of a myxoid cyst.

**Keywords:** Finger phalanges; Nails; Neoplasms; Synovial cyst

### INTRODUCTION

Giant cell tumor of the tendon sheath is a benign soft tissue (synovial membrane) tumor with onset between the third and fifth decades of life.<sup>1,2</sup> It is often located on volar aspects of fingers (two-thirds of cases), most commonly on the second and third fingers, near the distal interphalangeal joint.<sup>1</sup> Toe involvement is rare.<sup>3</sup> It usually presents as a painless slow-growing mass, which can mechanically interfere with the joint function. Adjacent skin is usually normal.<sup>1,3</sup> It rarely exceeds 5 cm in diameter.<sup>4</sup> Histologically, it is characterized by a population of different cells, including stromal round cells, multinucleated giant cells, and foam cells filled with lipids and hemosiderin deposits.<sup>5</sup> Although studies identified intracellular lipid deposits, no evidence of high cholesterol has been proven in these patients.<sup>5</sup> Radiology reveals soft tissue edema and even adjacent cortical erosion due to the pressure effect caused by the tumor.<sup>1</sup> Calcification is mentioned in 5.6%-16.7% of cases.<sup>6</sup>

Morphological classification divides giant cell tumors of the tendon sheath into two types: located nodular type (most commonly found on the hands and surrounded by pseudocapsules) and diffuse type (usually located around large joints). The diffuse type often presents as multicentric tumors or satellite lesions without encapsulation and is responsible for most cases of recurrence reported so far.<sup>5</sup> The localized form has a slight predilection for females, while the diffuse form affects both sexes equally.<sup>6</sup>

Despite its relatively frequent occurrence, giant cell tumors of the tendon sheath are rarely reported in the dermatologic literature, probably because they are mainly treated by hand surgeons.<sup>3</sup> In this paper, we present a case report of giant cell tumor of the tendon sheath clinically mimicking a myxoid cyst.

### CASE REPORT

We report a 49-year-old female patient presented with a painless, nodular, and translucent lesion near the distal interphalangeal joint of the third right finger. The tumor had a fibroelastic consistency with gradual growth for about two years. Adjacent skin revealed a mild erythema (Figure 1). On translucency examination, the lesion became more evident and well defined (Figure 2). Hand X-ray revealed no bone lesions. Based on the clinical findings, our initial diagnostic hypothesis was myxoid cyst, as it typically is located in the proximal nail fold in middle-aged women.<sup>7</sup> We performed an uneventful excision of the nailfold (Figure 3). Based on intraoperative findings, the tumor was classified as located nodular type. Histopathology revealed multinucleated giant cells surrounded by round cells and collagen rich hypocellular areas, as well as areas with xanthomatous histiocytes in surrounded by round cells, suggesting the diagnosis of tenosynovial giant cell tumor (Figures 4 and 5). The patient presented good postoperative evolution without signs of local recurrence to date (Figure 6).

### DISCUSSION

The etiology of giant cell tumor of the tendon sheath is not well defined. The hypothesis of a true neoplasm – resulting from sesamoid bones, from the synovial membrane, or from primitive mesenchymal cells – has been proposed. However, most authors favor the hypothesis that the condition is a reactive inflammation process.<sup>2</sup> The present case seems to contribute to the theory of a true neoplasm, since there was no report of local trauma or comorbidities.

Complete local excision is the treatment of choice for giant cell tumor of the tendon sheath<sup>1</sup>, but recurrence rate ranges from

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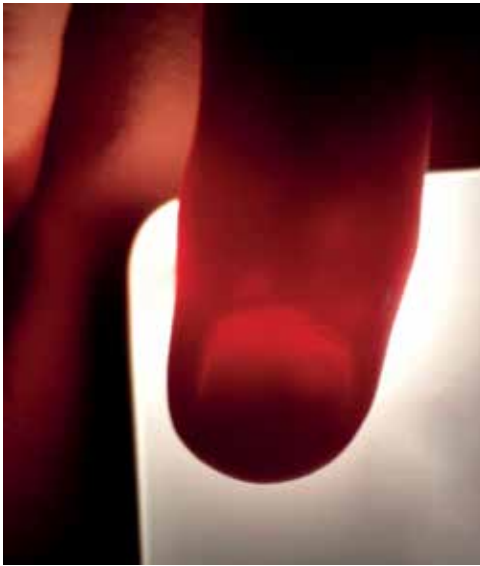
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<sup>1</sup> Department of Dermatology of the Irmandade Santa Casa de Misericórdia de Porto Alegre – Porto Alegre (RS), Brazil.

<sup>2</sup> Department of Pathology of the Irmandade Santa Casa de Misericórdia de Porto Alegre – Porto Alegre (RS), Brazil.



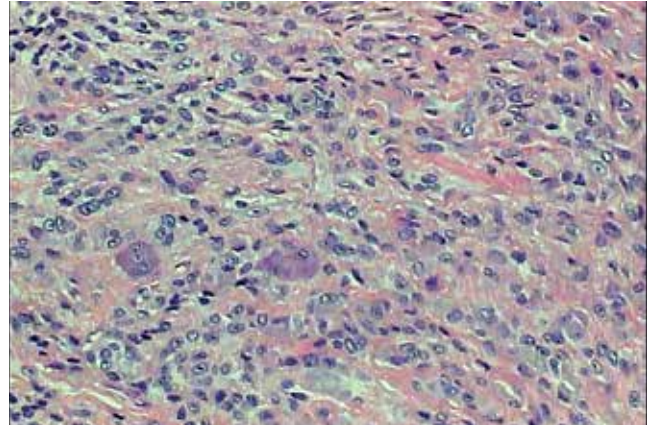
**Figure 1:** Lesion with nodular aspect next to the proximal nail fold on the third right finger. Convexity and depressions on the plate



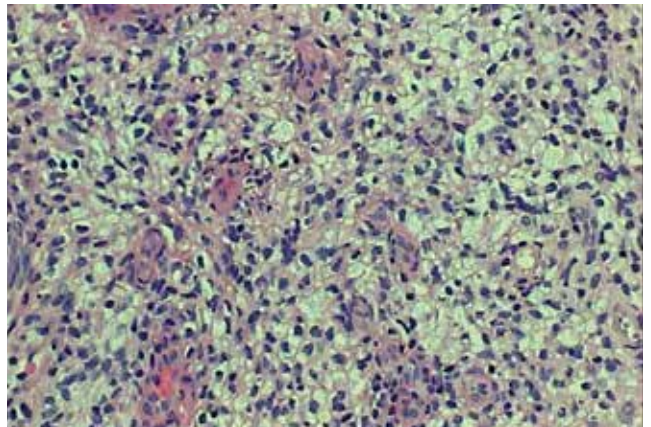
**Figure 2:** On translucency examination, presence of translucent circular areas forming halos



**Figure 3:** Surgical exploration revealed multilobulated tumor with smooth and shiny surface in xanthomatous tissue



**Figure 4:** Histiocytic proliferation forming giant cells similar to osteoclasts. Presence of macrophages, collagen, and hyaline fibrous tissue (Hematoxylin & eosin x10)



**Figure 5:** Xanthomatous histiocytic differentiation. Proliferation of histiocytes and fibroblasts in xanthomatous areas (Hematoxylin & eosin x10)



**Figure 6:** Satisfactory postoperative evolution without signs of recurrence after two months

4%-44%, as described in the literature.<sup>5</sup> The risk of recurrence is associated mainly to incomplete resection or other reasons, such as anatomical location (distal interphalangeal joints or thumb interphalangeal joint), presence of radiography erosion, association with

degenerative disease, and diffuse subtype.<sup>5,8</sup> Age, gender, lesion size, and volar location seem to have no influence on recurrence, which probably occurs due to the lobular nature of the tumor with hidden extension around the tendon sheath.<sup>3</sup> All surrounding tissues shall be examined for satellite lesions and their connections, which must be removed with the aid of a surgical microscope or a magnifying glass. Local irradiation has been applied as an adjuvant therapy to prevent recurrence.<sup>2</sup>

Differential diagnosis of hand tumors include lipomas, hemangioma, foreign body, myxoid cyst, synovial carcinoma, tophaceous gout, glomus tumor, tuberosus osteitis, epidermal cyst, fibroma, and metastasis.<sup>1,3</sup> Granuloma annulare and erythema elevatum diutinum should also be considered.<sup>3</sup> Mass aspiration can help rule out tophaceous gout, but the definitive diagnosis is confirmed only with surgical excision and histopathological examination of the lesion.<sup>8</sup> Only 20% to 30% of giant cell tumors of the

tendon sheath are clinically diagnosed before surgery.<sup>3</sup> Thus, histopathology is virtually important, since the lesion can mimic other medical conditions, which often have different types of treatment, as reported in our case.

Although the condition is routinely benign, malignant degeneration has been reported in exceptional cases.<sup>3</sup> The diffuse type can be locally aggressive, with reports of possible multiple recurrence and malignant transformation.<sup>4</sup> These data reinforce the need for early and accurate diagnosis, and proper treatment.

Although giant cell tumor of the tendon sheath is little reported by dermatologists, its recognition is essential since it is a tumor of still uncertain origin and behavior, which may mimic other diseases. The reported case allows a reflection on the differential diagnosis of hand tumors and the need for histopathology and excisional treatment when appropriate.□

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## MAILING ADDRESS:

Renan Minotto  
Rua Comendador Rheingantz, 750 apto. 701  
Bela Vista  
90450-020 Porto Alegre, RS  
Brazil  
E-mail: rminotto@gmail.com

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