Superficial Acral Fibromyxoma, Appearing as Retronychia: A Rare Clinical Entity

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Established Facts

- Retronychia is a disorder caused by ingrowth of the proximal nail plate into the proximal nail fold and is suspected in cases of unresolved chronic proximal paronychia.
- Superficial acral fibromyxoma is a rare benign tumor of soft tissues commonly located in the subungual or periungual area.

Novel Insights

- Total avulsion of the nail constitutes the treatment of choice for retronychia and can reveal the existence of a subungual superficial acral fibromyxoma as a cause of this condition.

Keywords

Immunohistochemistry · Retronychia · Superficial acral fibromyxoma · Surgical excision

Abstract

We report a case of a subungual superficial acral fibromyxoma (SAFM) in a 37-year-old male patient who presented with a persistent chronic proximal paronychia of the big toenail of the right leg. Our clinical diagnosis was retronychia, an often misinterpreted condition, which must be suspected in cases of persistent paronychia, especially in the setting of trauma. The nail plate avulsion revealed a subungual tumor, which was surgically excised. Histopathology in combination with immunohistochemistry revealed features suggestive of SAFM. The follow-up examination of the patient showed no recurrence 9 months after the surgery.

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Introduction

Superficial acral fibromyxoma (SAFM) is a rare mesenchymal tumor of soft tissues with a predilection for the subungual or periungual region, which was initially described by Fetch et al. [1]. It generally presents as a slow-growing painless nodule in middle-aged adults [1, 2]. SAFM has a benign behavior, but it may show persistence or recurrence if not properly treated [3]. Its differential diagnoses include other neoplasms, such as dermatofibrosarcoma protuberans and myxoid neurofibroma [2].

Case Report

We present the case of a 37-year-old male patient with a personal history of cystic fibrosis and diabetes mellitus Type I who was referred to our department due to a chronic proximal paronychia

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Fig. 1. Xanthonychia, onycholysis, thickening of the proximal portion of the nail plate, and proximal paronychia.

Fig. 2. Resection of the lesion en bloc, under tourniquet control.

Fig. 3. Normal regrowth of the nail plate, 9 months after the surgery.

of the big toenail of his right leg, unresolved after systemic antifungal and antibiotic therapy. The patient reported chronic trauma at the site of the nail apparatus.

Physical examination revealed xanthonychia, onycholysis, thickening of the proximal portion of the nail plate, and interruption of nail growth (Fig. 1). The clinical diagnosis was retronychia, and initially, topical therapy with clobetasol propionate 0.05% (an ultrapotent class I corticosteroid) under occlusion was prescribed for 3 months. As no improvement was noted, total ablation of the nail plate was warranted.

During the surgical procedure, the nail plate was detached from the nail fold, under distal anesthetic block. However, bulging of the lunula was observed and a transverse incision of the distal matrix part was performed, revealing a pink-to-flesh-colored well-demarcated lesion. Therefore, we proceeded to a resection of the lesion en bloc under tourniquet control and sent it for histological examination (Fig. 2).

Histological results showed proliferation of fibroblast-like cells on myxoid stroma, with rare nuclear atypia. A rich vascular network was noticeable. Immunohistochemical analysis showed CD34 positivity and negativity for neural and muscular differentiation markers (S-100, SMA, EMA, and desmin). These histological results were compatible with the diagnosis of SAFM. After 9 months, the patient is asymptomatic, without signs of tumor recurrence, while the nail plate appears to regrow normally (Fig. 3).

Discussion

We present herein a case of a subungual SAFM, which appeared clinically as retronychia. Retronychia refers to the proximal nail plate ingrowth into the proximal nail fold [4]. Approximately 140 cases of retronychia have been reported in the literature until now [5–7], whereas recently a large series of 56 cases was published [6]. The diagnosis of retronychia is clinical and must be suspected in cases of persistent paronychia, particularly in the setting of trauma.

Chronic proximal paronychia, onycholysis, emerging of granulation tissue from under the nail fold, xanthonychia, and thickening of the proximal portion of the nail plate include the clinical diagnostic criteria for retronychia [7]. The differential diagnoses of retronychia include subungual benign and malignant tumors, cysts, and diseases that can present as chronic paronychia (e.g., fungal and bacterial infections) [7]. Total avulsion of the nail not only constitutes the treatment of choice for retronychia but also confirms the diagnosis and allows rapid pain relief. In our case, avulsion of the nail revealed the existence of a subungual mass, suggestive of SAFM.

SAFM is a relatively rare benign neoplasm commonly located in the acral extremities, especially in the fingers and toes, and presents as a nodular, slow-growing, painless mass. A total of 314 cases of SAFM are found in the literature [8], whereas only a few cases have been reported as trauma induced. The lesion affects usually young adults, with a higher frequency in males than females. Radiologic examination rarely shows bone alterations [9].

Histologically, SAFM is composed of proliferation of fusiform fibroblasts inside a myxoid, myxocollagenous, or collagenous matrix. A rich microvascular network and local infiltration of mast cells can be seen in the tumor [1, 3, 10]. Immunohistochemistry is recommended for the diagnosis and shows CD34, CD99, and EMA positivity, while the results for neural and muscular markers (S-100, HMB-45, SMA, desmin, and actin) are negative [1, 10, 11].

Because of the rarity of this clinical entity, awareness is required. In the differential diagnoses of SAFM are included not only CD34-immunonegative tumors, such as glomus tumor, sclerosing perineuroma, acquired digital fibrokeratoma, and periungual fibroma, but also CD34-immunopositive tumors, such as superficial angiomyo-

ma, dermatofibrosarcoma protuberans, and myxoid neurofibroma [2, 9].

Gross total resection of the lesion is the gold standard of treatment. However, it may recur locally in approximately 20–25% of the cases, when initial margins are positive [12]. Furthermore, malignant transformation or metastasis of SAFM has not been documented in the literature [12].

To our knowledge, this is the first case of a subungual SAFM which appeared clinically as retronychia. Retronychia might be the result of different conditions such as chronic trauma or subungual tumors. Therefore, when we face a retronychia resistant to potent/ultrapotent topical steroids (class I and II) under occlusion, surgical ablation is warranted. Additional total avulsion of the nail remains the treatment of choice for retronychia and can reveal the existence of a subungual tumor as a cause of this condition, as in our case.

Statement of Ethics

The patient's consent was obtained for the publication of Figures 1–3 and details of the case.

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Author Contributions

Evangelia Kalloniati: contributed to the design of this work, took the patient's consent for the publication of the case, and co-wrote the manuscript. Stella Arampatzi: co-wrote the manuscript. Christina Ouzouni: was responsible for the collection of the figures of the patient before and after the surgery and revised the manuscript for important intellectual content. Despoina Papathemeli: helped during the surgical procedure and supervised the accuracy of any part of this work. Elizabeth Lazaridou: supervised the management of the case and the writing of the manuscript. Myrto-Georgia Trakatelli: was responsible for the management of the case, performed the total ablation of the nail plate, and supervised the writing of the manuscript.