DOI: 10.15386/cjmed-517

CLINICO-PATHOLOGIC, DERMOSCOPIC AND ULTRASOUND EXAMINATION OF A RARE ACRAL TUMOUR INVOLVING THE NAIL - CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract

There is a large spectrum of tumors presenting as nodular lesions that may affect the subungual space. We report the case of a 62-year-old woman presenting with a rapidly growing nodular lesion under the nail of the first left toe. Non-invasive examinations using dermoscopy, ultrasonography and elastography were performed for the preoperative assessment of the lesion. The biopsy of the lesion revealed superficial acral fibromyxoma, a benign tumor with predisposition for acral sites. The patient underwent radical surgery with wide resection margins. This is the first case report of a superficial acral fibromyxoma affecting the subungual region characterized by dermoscopic, ultrasonographic and elastographic features. We also performed a short review of the literature.

Keywords: superficial acral fibromyxoma, dermoscopy, ultrasonography, elastography

Introduction

Superficial acral fibromyxoma (SAF) is a rare benign tumor that appears frequently in the fifth decade of adult life. It is a soft-tissue tumor often reported in the subungual and periungual regions of fingers and toes [1,2]. It was recognized as a distinct clinical and pathological entity after Fetsch et al. [3] published a clinico-pathological and immunohistochemical analysis of 37 patients presenting a special soft-tissue tumor with predilection for fingers and toes.

The diagnosis and treatment of nail unit tumors always represent a challenge. The technical difficulties of a nail unit biopsy, the painful procedure and the patient's concern for permanent nail dystrophy subsequent to this procedure, place the doctors in a difficult position.

Because subungual tumors do not have specific clinical manifestations, imaging represents an important step in evaluation and differentiation of these tumors. Dermoscopy allows in vivo evaluation of different parts of the nail apparatus, being able to describe colors and its microstructure [4]. Radiography can detect calcification and it can also evaluate adjacent bone structures. High frequency ultrasonography (HFUS) and color Doppler imaging are effective methods to evaluate tumors, but are limited in tissue characterization. The tumoral detection can be impeded by various artefacts, small tumor size, and also flat tumor shape [5]. Elastography is also a non-invasive method that allows *in vivo* evaluation of soft-

Manuscript received: 26.07.2015
Accepted: 17.08.2015
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tissue elasticity, being able to increase the specificity of US in assessment of tumors [6].

In this case report we discuss and illustrate a rare acral tumor involving the nail apparatus, and correlate for the first time the dermoscopic and ultrasonographic features with clinical and pathological findings. We also performed a short review of the literature regarding SAF.

Case report

A 62 year old woman referred to the Dermatology Department with a 4 months history of enlarging subungual mass on the first left toe. She reported repeated trauma, pain when she wore closed shoes and pigmentation nail changes over the last year. Physical examination of the toe nail revealed a 15 mm, firm, painful, reddish- yellow nodule that destroyed the distal nail plate, partial yellow-brown nail discoloration, and thickening of the nail (Figure 1a). Dermoscopy (DermLite DL3N- 3Gen USA) revealed structureless brown discoloration, structureless homogeneous red, whitish and yellow areas (Figure 1b). The direct microscopic examination showed the presence of fungi. B-mode gray ultrasonography (US) showed a nodular, hypoechoic, inhomogeneous, 12.7/20.2 mm structural lesion that distorted the nail and presented a cranial hypoechoic extension (Figure 2a). On Color Doppler examination the lesion presented variable vascularity and necrotic zones (Figure 2b). Strain elastography showed variations in hardness with intermediate rigidity predominating (Figure 2c). A diagnostic biopsy was performed and revealed a polypoid, ulcerated, and well vascularised neoplastic dermal tumor (Figure 3a). A proliferation of fusiform cells immersed in a myxoid stroma with no relevant atypia was described (Figure 3b). Immunohistochemical (IHC) study reported positive results for CD34 (Figure 4a), CD99 (Figure 4b), CD10 (Figure 4c) and was negative for EMA. These findings led to the diagnosis of superficial acral fibromyxoma. The therapeutic approach included complete removal of the tumour, as well as the nail, with wide resection margins in order to avoid recurrence. After a 12 months follow-up there was no recurrence of the tumor.

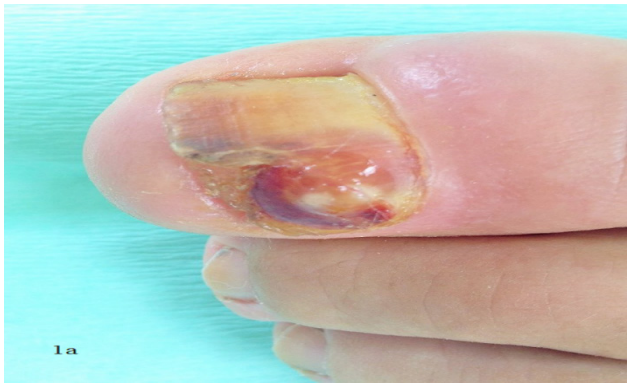

Figure 1a. Clinical picture: subungual nodular tumor.

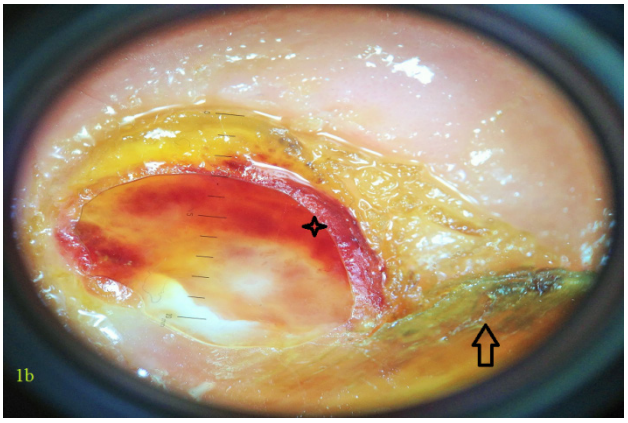

Figure 1b. Dermoscopy: star-structureless homogeneous red area within the tumor: arrow - structureless brown discoloration of the nail.

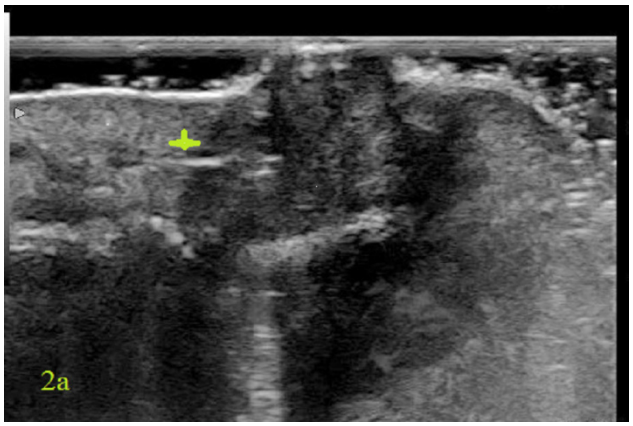

Figure 2a. B-mode gray US: nodular, hypoechoic, inhomogeneous tumour that distorts the nail. Star-cranial hypoechoic extension.

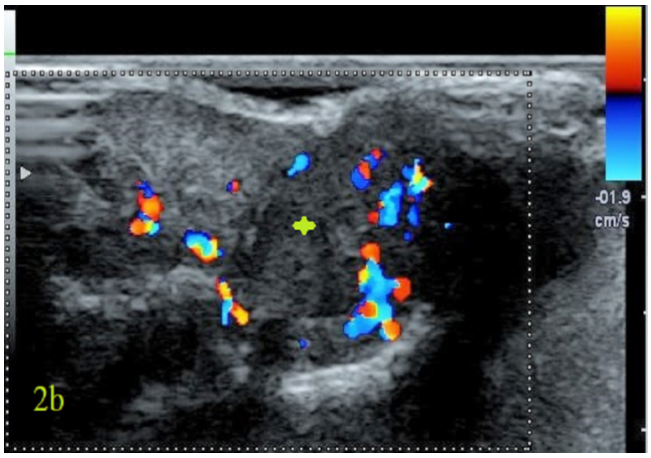

Figure 2b. Color Doppler: variable vascularity. Star- necrotic zones.

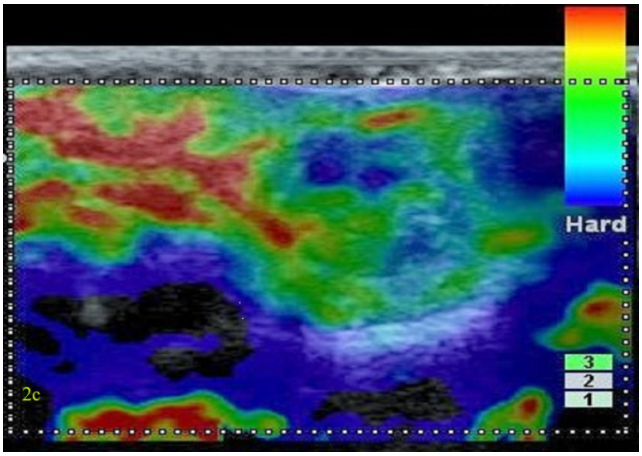

Figure 2c. Strain elastography: intermediate rigidity.

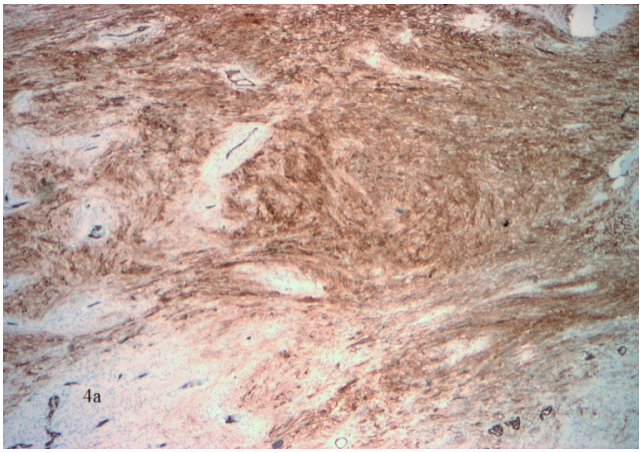

Figure 4a. IHC staining: CD34 diffuse expression 2 x.

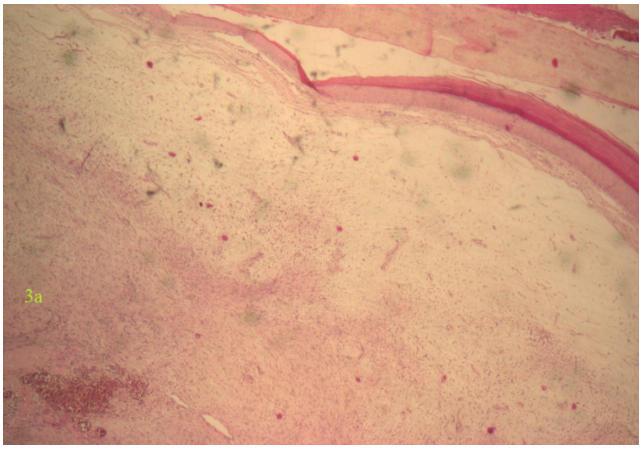

Figure 3a. Histhopathology of SAF 2 x HE: nonencapsulated, well-circumscribed tumor of the dermis.

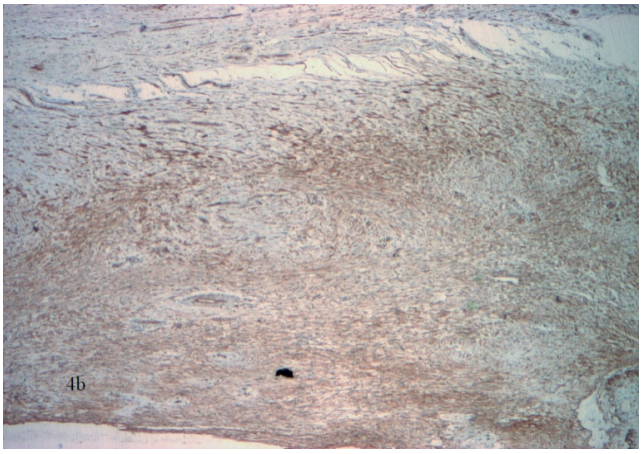

Figure 4b. IHC staining: CD99 diffuse expression 2 x.

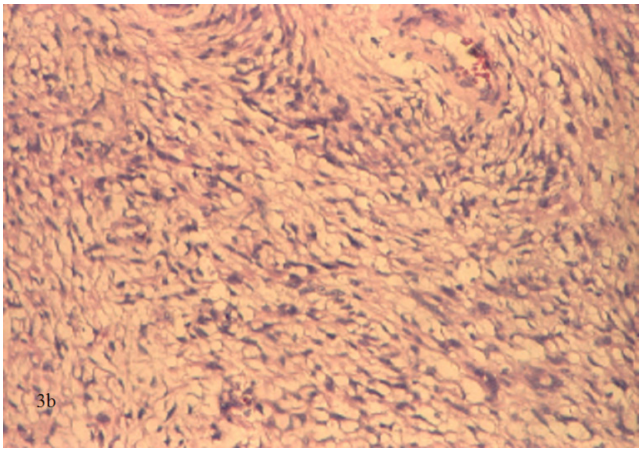

Figure 3b. Histopathology of SAF 10 x HE: a proliferation of fusiform cells immersed in a myxoid stroma with no relevant atypia.

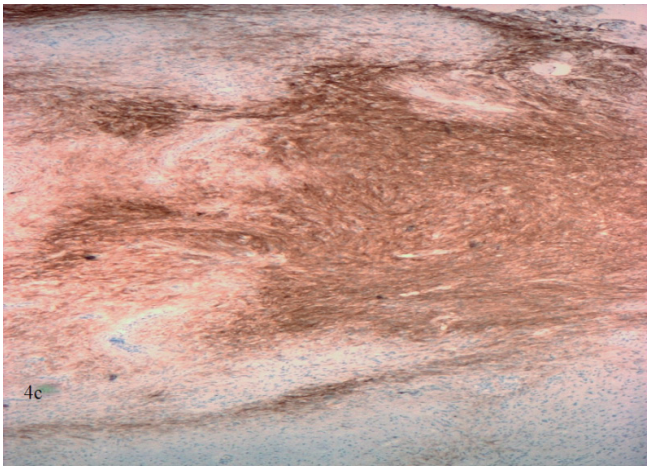

Figure 4c. IHC staining: CD10 diffuse expression 2 x.

Discussion

SAF is a nodular soft-tissue tumor, of solitary presentation and various consistency, from gelatinous to firm [1,2]. Different clinical presentations were reported varying in shape and color: flesh-colored, white, pink, red nodular mass with a dome, verrucous or polypoid shape [1]. Studies usually report a slow-growing tumor that varies in size from a few mm to several cm in diameter and can appear months to years prior to evaluation [1,7]. SAF has a predilection for male sex with a sex ratio estimated as 2:1 [8]. From almost 260 cases of SAF reported in literature only five cases had sub-matricial presentation: three had solely submatricial localization and in two cases there was a secondary sub-matricial extension of a SAF affecting the nail bed [9].

The particularity of our case is the appearance of the lesion in a woman, as a firm, painful, nodular mass with a reddish-yellow appearance, affecting the nail bed with a sub-matricial extension identified only in the B-mode US.

Dermoscopy is a very useful tool in the evaluation of pigment or vessels within a nail lesion, but is unable to evaluate the inner structure of the nail bed [4]. Dermoscopically, the patient's nail presented with a structureless yellow-brown discoloration and partial nail plate destruction which are characteristic of a fungal infection [4]. The nodular nail bed lesion showed structureless homogeneous red areas (Figure 1 b), which usually appear in vascular lesions or in amelanotic/hypomelanotic melanoma, associated with structureless, homogeneous whitish and yellow areas which are characteristic of squamous lesions [4]. Our findings suggest that subungual SAF has nonspecific dermoscopic characteristics, but larger studies are needed to confirm this hypothesis.

Some studies have showed that HFUS completed by color Doppler examination of subungual tumors provides useful information about tumor size, location and shape of the tumour and also the internal characteristics: vascular, cystic, solid or mixed. It is also important in monitoring the lesions [5]. Subungual tumors present in US as hypoechoic, homogeneous or heterogeneous masses. Color Doppler US can appreciate tumor vascularization, the blood flow amount, the size and the distribution of vessels within the tumor adding important information for the final diagnosis [5]. In our case the US features were concordant with those described in the literature, but we found no correlation between the dermoscopic and US appearance of vessels. For a better evaluation we decided to perform elastography, which has an important role in evaluation of extra cutaneous tumors, but has no established role in the assessment of nail tumors [10]. Our tumor showed intermediate rigidity in elastography being the first case reported in the literature of subungual SAF examined by elastography (Figure 2c) which was useful in evaluation.

Although it is postulated that benign tumors respect the general architecture of the nail, up to 96% of toe tumors involve and can destroy the nail architecture [2,11]. There are studies who reported erosions or lytic lesions of underlying bones in a third of cases [2]. Our case revealed no visible bone alterations on radiography.

The histopathology of SAF is characterized by the presence of a non encapsulated, well-circumscribed tumor of the dermis sometimes with extension into the subcutaneous tissue, fascia or periosteal layer [1]. The tumor contains stellate or spindle-shaped fibroblast-like cells which are arranged in a storiform or fascicular growth pattern with a myxoid, myxocollagenous or predominantly collagenous surrounding stroma [2]. The presence of mast cells in an increased number and an accentuated vasculature can be observed [2,11]. Mitotic figures are rare, and nuclear atypia is slight or absent [12]. The IHC profile of SAF is characterized by the presence of diffuse or focal expression of CD34 in up to 90% of cases [11]. CD99, CD10, EMA and nestin immunoreactivity can also be focally positive. Staining for S-100, HMB-45, SMA, desmin, actin and cytokeratin is negative [1-3,7,8]. The IHC study in our case showed positive results for CD34, CD99 and CD10 and was negative for EMA (Figure 4a,b,c).

The standard treatment of SAF is surgery with complete local resection of the tumor. There were reports of cases treated by partial excision, and partial / complete digital amputation [1]. The incomplete tumoral excision can lead to persistent or recurrent cases in up to 20% of cases [1,2]. In our case the radical surgery with wide resection margins lead to no recurrence after 12 months of follow-up.

This is the first reported case in the literature of subungual SAF described clinically, dermoscopically and by US examination (B-mode US, colour Doppler, strain elastography).

Acknowledgement

"This paper was published under the frame of European Social Found, Human Resources Development Operational Programme 2007-2013, project no. POSDRU/159/1.5/S/138776".

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