CD34 negative superficial acral fibromyxoma: A rare case report

ABSTRACT

Superficial acral fibromyxoma (SAF) is a slow growing soft tissue tumor that mainly appears in the acral areas. Here, we report a case of a SAF with distinctive immunophenotype charachteristics. An 18-year-old female was referred to our clinic with the complaint of painless subungual nodule of great toe for a few months. The diagnosis of SAF was made according to histopathology and immunohistochemical (IHC) study, however, the IHC assessment showed positive staining with vimentin, focal reaction with smooth muscle actin, negative reaction with CD34, and positive staining pattern with CD99. These IHC findings are unusual for SAF. This reported case of SAF supports the fact that, although CD34 expression is characteristic for SAF, it is not always present.

Key Words: *Benign soft tissue tumor, CD34, immunohistochemical (IHC) study, superficial acral fibromyxoma*

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Introduction

Superficial acral fibromyxoma (SAF) is a slowly growing soft tissue tumor that tends to appear in the acral sites. First described in 2001, Fetsch *et al*., reported its clinicopathologic features and immunohistochemistry (IHC) findings, and since then, around 170 cases have been reported.[1] There have been quite a few reports of cases with this myxoid tumor.[2-5] Clinically, it appears as a slow growing, well-circumscribed neoplasm of fingers and toes in middle-aged adults. Pathological findings include a dermal or subcutaneous tumor with spindled and stellate-shaped cells that are embedded in myxoid or collagenous matrix.[1] It stains positive for CD34 and focally positive for epithelial membrane antigen (EMA) and CD99. There have been a few reports of cases that were negative for CD34, however, to our knowledge, none of the cases reported before showed the distinct immunophenotype seen in our case.[1] Herein, we report a case of CD34 negative SAF with clinical pictures and IHC investigation.

Case Report

An 18-year-old female presented to our dermatology clinic with the complaint of a slowly growing subungual nodule since a few months. The lesion was painless, she did not mention any history of trauma in the area, and the past medical history was negative for any relevant disease. The physical examination showed a nontender subungual, nodular lesion with 12 mm diameter in the right big toe [Figure 1a]. The lesion caused mild deformity of the nail. Radiological examination did not show any changes in the underlying bone.

Punch biopsy of the lesion revealed a fairly circumscribed dermal proliferation of spindled and stellate cells with loose storiform and fascicular pattern embedded in fibrocollagenous stroma with foci of myxoid change [Figure 2a and b]. The vasculature was prominent and mitotic activity and nuclear atypia were absent. The lesion was completely excised by a plastic surgeon with free margin and did not recur in 6 months follow-up [Figure 1b]. IHC staining of our case revealed positive staining with vimentin, focal reaction with smooth muscle actin, negative reaction with CD34, and positive staining pattern with CD99 [Figure 3a-d]. The IHC for S-100 was also performed and turned out to be negative. The diagnosis of SAF was made according to the histopathology and IHC data.

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Discussion

SAF is a relatively rare dermal or subcutaneous myxoid tumor, which is well-circumscribed and unencapsulated. It tends to be slow growing, and painless, with a male preponderance.[6] The size of the lesions could be between 0.6 to 5.0 cm (mean: 1.75 cm). In a series of cases with SAF, the lesion existed from 3 months to 30 years (median duration: Approximately 3 years) prior to treatment.[1] It habitually arises on the fingers and toes of middle-aged adults but unusual sites of occurrence such as heels have also been reported.[7]

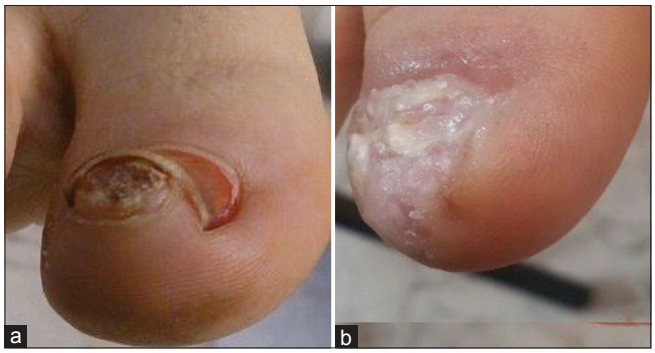

Figure 1: (a) Nontender subungual, nodular lesion in the right toe; (b) after excision of the lesion

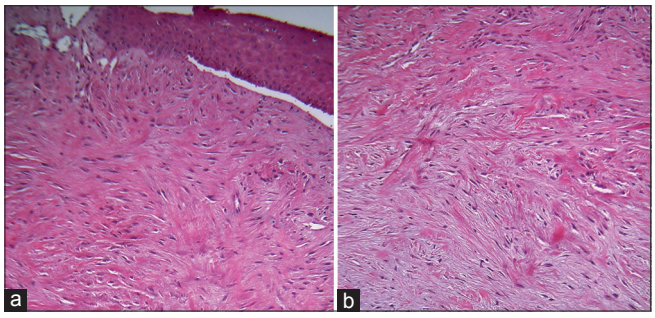

Figure 2: (a and b) High power view of bland spindle cells in a fibromyxoid stroma. Note the normal looking epidermis overlying the tumor (H and E ×40)

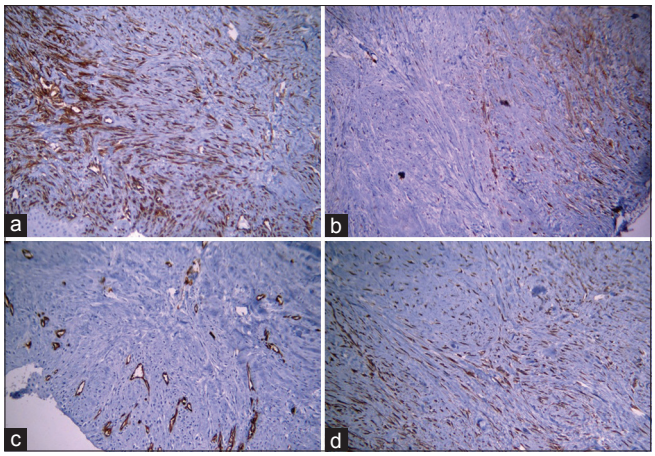

Figure 3: Immunohistochemical (IHC) study; low power view of neoplastic spindle cells which show IHC positive staining with vimentin (a), focal reaction with smooth muscle actin (b), negative reaction with CD34 (c), and positive staining pattern with CD99 (d). (IHC ×40)

Adjacent nail might show hyperkeratosis or onycholysis occasionally accompanied by pain on compression. Erosion of the underlying bone is rare but it has been reported.[8] The frequent deformity of the nail plate necessitates the removal of the nail plate during surgical procedure.[1,6,9] Our case did not show involvement of the underlying bone, however, the nail bed was involved and the nail unit was completely excised during the surgical procedure.

SAF is a benign neoplasm and there have been no reports of malignant transformation or metastasis; however, there are a number of recurrent cases that have been related to incomplete resection.[7,8]

Histological appearance of SAF is more or less consistent with virtually all tumors presenting with spindle cells. It shows an indistinct storiform and fascicular pattern embedded in a myxoid/fibromyxoid/collagenous stroma often with mildly accentuated vasculature and increased numbers of mast cells. Atypia and mitotic figures are generally absent; however, there are reports of cells with atypical features found in this tumor.[7]

Neoplastic cells in SAF usually present with immunoreactivity for CD34, CD99, and EMA and negative staining for actin, desmin, keratins, S100 protein, and HMB45.[1-4,6] Variation in IHC staining is observed in cases with SAF. Our case exhibited positive staining for CD99, vimentin and SMA and negative staining for CD34, and S100. Immunoreactivity to CD34 is a common feature of SAF; however, there are tumors with negative staining for this marker.[3,5] In fact, in a detailed characterization of 124 cases with SAF, approximately one-third of lesions were negative for CD34.[10] Therefore, a diagnosis of SAF should be considered even in the absence of reactivity for CD34.

SAF should be differentiated from both benign and malignant myxoid lesions. Benign lesions include those with proliferation of spindle cells and myxoid lesions with spindle-shaped cells such as myxoid neurofibroma, superficial angiomyxoma. Myxoid neurofibroma can be excluded by negative staining for S-100.[11] Angiomyxoma stains positive for CD34 but has a predilection for head, neck, and trunk and has prominent hyalinized vessels.[12]

Malignant myxoid lesions include myxoid dermatofibrosarcoma protuberans, acral myxo-inflammatory fibroblastic sarcoma, and low-grade fibromyxoid sarcoma. Focal storiform pattern in this tumour should be differentiated with dermatofibrosarcoma protuberans, which shows CD34 reaction but occurs very rarely in acral sites.

Acral myxoinflammatory fibroblastic sarcoma has a predilection for subcutaneous soft tissues of extremities and

shows prominent inflammatory cell component and bizarre tumor cells and CD34 is diffusely positive in most cases.[13]

The other differential diagnosis of our case is onychomatricoma in which there are similar stromal cells, however, the epidermal changes in onychomatricoma makes it quite different.[14] Glomus tumor is another differential diagnosis which is almost always painful and usually stains positive for vimentin and smooth muscle antigen and negative for desmin and CD99. CD34 can also be positive in glomus tumor.[15] Since this tumor was CD99 positive, it should be distinguished from monophasic synovial sarcoma, which usually shows areas of calcification and at least focal reaction to cytokeratin.[16]

In conclusion, SAF seems to be a rare subungual tumor with various IHC properties. A diversity of lesions can occur in the subungual area, and proper diagnosis and treatment of these lesions requires a meticulous histopathological and IHC evaluation.

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Conflicts of interest

There are no conflicts of interest.

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