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 was referred to our clinic with the complaint of painless subungual nodule in the great toe for a few months. 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.
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. 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. It habitually arises on the fingers and toes of middle-aged adults but unusual sites of occurrence such as heels have also been reported.

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. The frequent deformity of the nail plate necessitates the removal of the nail plate during surgical procedure. 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. Angiomyxoma stains positive for CD34 but has a predilection for head, neck, and trunk and has prominent hyalinized vessels.

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. 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.

Neoplastic cells in SAF usually present with immunoreactivity for CD34, CD99, and EMA and negative staining for actin, desmin, keratins, S100 protein, and HMB45. 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. In fact, in a detailed characterization of 124 cases with SAF, approximately one-third of lesions were negative for CD34. Therefore, a diagnosis of SAF should be considered even in the absence of reactivity for CD34.

Malignant myxoid lesions include myxoid dermatofibrosarcoma protuberans, acral myxoinflammatory 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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