

Case Report

Superficial Acral Fibromyxoma: a Rare Tumor Diagnosed by Cytology

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ABSTRACT

Superficial acral fibromyxoma (SAFM) are rare soft tissue tumors predominantly involving hands and feet of adults. There are only a few reported cases of this benign neoplasm, but no fine needle aspiration cytology (FNAC) diagnosis of this tumor is reported in the literature. A 48 year old female presented with a 5 × 3.5 cm globular, firm swelling over the right fifth toe. FNAC of the lesion was interpreted as superficial acral fibromyxoma. Subsequent histopathology confirmed the diagnosis and demonstrated cytohistological correlation for the first reported time in the literature. SAFM may recur if not adequately excised. Preoperative FNAC diagnosis will help the surgeons to plan for wider excision to prevent recurrence.

Keywords: Fibromyxoma, Cytology

Introduction

Superficial acral fibromyxoma (SAFM) is a recently described distinct clinicopathological entity. Fetsch *et al.* presented the largest series of 37 cases in 2001 and coined the term (1). Till to date, only very few case reports describing typical histological features of SAFM have been published (2-4).

SAFM is a rare, benign soft tissue neoplasm involving predominantly fingers and toes with a strong predilection for the great toes. This tumor usually presents as a slow growing tender mass in adult males (5). The commonest location is subungual or periungual (1, 3). Due to location, these tumors may cause nail plate deformities requiring removal during excision (1, 6). SAFM affects a wide age range 14-75 years with a

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slight male preponderance (1). Histologically, it is a poorly circumscribed mass mainly composed of spindle or stellate neoplastic cells that show a variable degree of pleomorphism in a myxocollagenous stroma (5). However, there is no reported case of FNAC diagnosis of SAFM in literature. We report a case of SAFM primarily diagnosed by FNAC and confirmed by histopathology later.

Case Report

A 48 year female presented to the FNAC Clinic of Bankura Sammilani Medical College, Bankura with a globular swelling over the subungual region of the fifth toe. The lesion had been present for 3 years. The lesion was painless, soft to firm and mobile. Aspiration yielded mucoid material. Smears were moderately cellular consisting of benign spindle cells in loose clusters and singly with fragments of thin walled

capillaries in a myxoid background (Fig. 1). No evidence of malignancy was noted. A diagnosis of superficial acral fibromyxoma was rendered with a recommendation for wide excision of the lesion.

Excisional biopsy was performed and the tissue was sent for histopathological examination. Gross examination revealed an oval, circumscribed, nonencapsulated solid tumor, oval in shape measuring (4.5×3×3 cm).

Cut section was solid, grey-white and fasciculataed. Histopathology demonstrated a moderately cellular tumor composed of fascicles of spindle and stellate cells arranged in a vague storiform pattern. Pleomorphism, mitotic figures and giant cells were not noted. Few mast cells were found in the tumor. The background was myxoid with many blood vessels present. A diagnosis of superficial acral fibromyxoma was confirmed (Fig. 2 and 3).

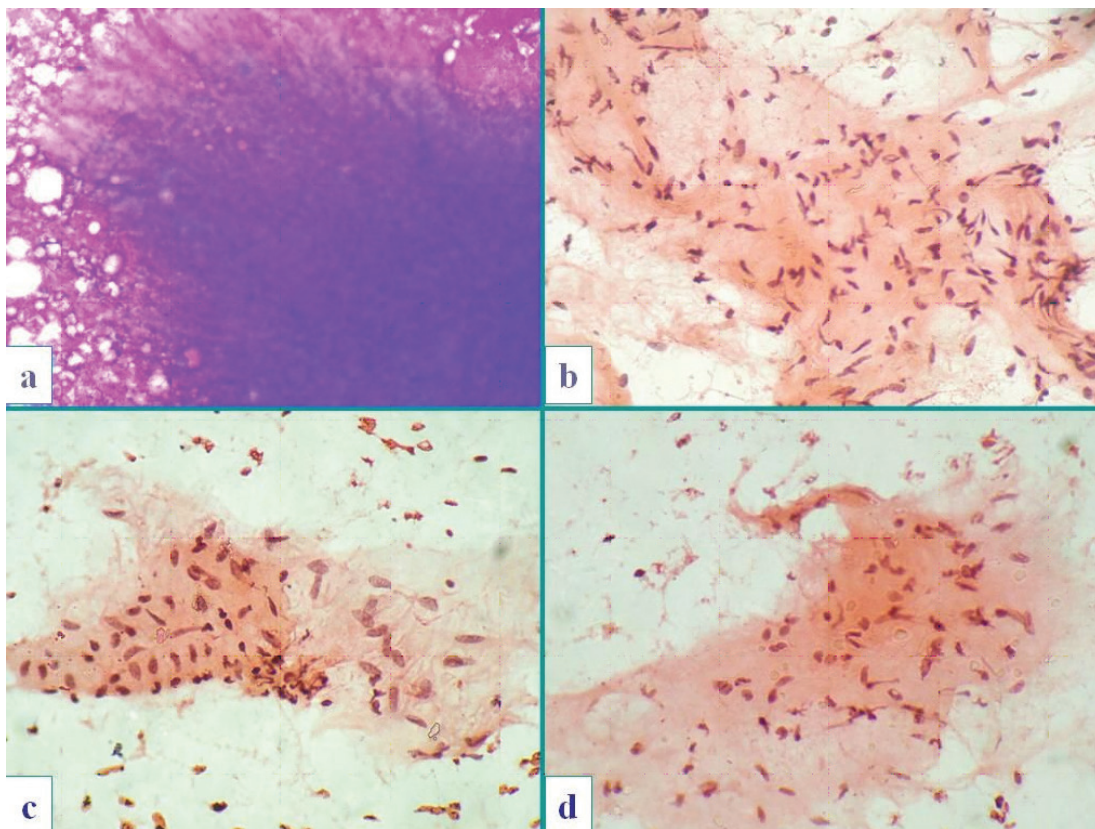


Fig. 1- FNAC smears show: a) benign spindle cells in loose clusters in a background of myxoid material. (MGG, ×100). b, c, d) loose clusters of spindle cells and stellate cells embedded in a fibromyxoid matrix. (Pap, ×400)

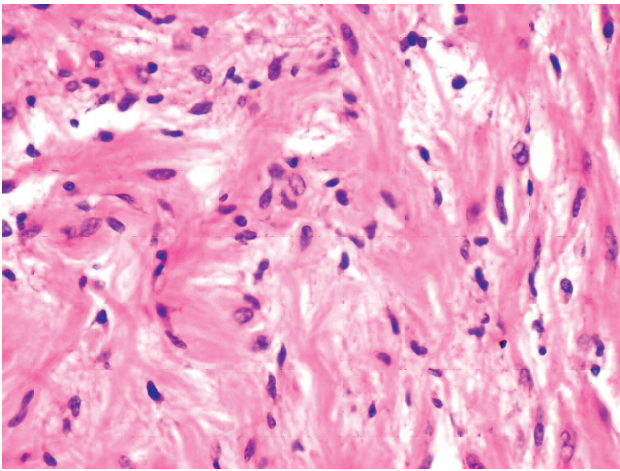


Fig. 2- Section shows proliferating spindle and stellate cells with storiform and fascicular patterns, embedded in a myxocollagenous stroma. (H&E, ×400)

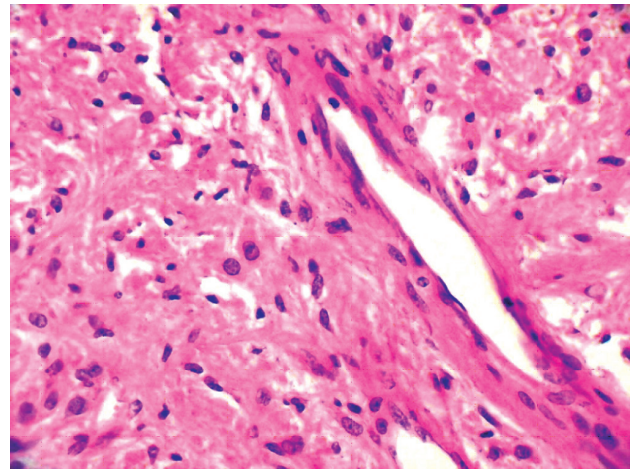


Fig. 3- Section shows blunt looking spindle cells in a loose storiform to fascicular pattern, increased number of blood vessels in a collagenous stroma with marked myxoid changes. (H&E, ×400)

Discussion

SAFM is a rare, distinctive soft tissue tumor with a predilection for the ungual region of the fingers and toes in middle-aged adults (7). This tumor was first described by Fetsch *et al.* in 2001 (1). For correct interpretation of SAFM, correlation of clinical, microscopic and immunohistochemical findings are important (1, 8). Immunohistochemical staining of SAFM is characteristic, being- positive for CD-34, CD-99, EMA, and CD10 and negative for S-100 protein (1).

The differential diagnosis for SAFM includes myxoid neurofibroma, fibroma of tendon sheath, glomus tumor, superficial angiomyxoma, acral fibrokeratoma, sclerosing perineuroma, cutaneous myxoma, myxoid fibrous histiocytoma and low grade myxofibrosarcoma.

Myxoid neurofibroma has a neural appearance with no increase in vascularity and S100 positive. Here immunoreactivity to S100 as a marker for neural origin and lack of vasculature in the tumor mass distinguish them from SAFMs (9).

Fibroma of tendon sheath is usually well-circumscribed and attached to a tendon sheath.

In contrast to a moderately cellular proliferation of spindled fibroblast-like and stellate cells seen in SAFM, this tumor shows sparse stellate cells embedded in a fibrocollagenous matrix with dilated or slit like vascular channels (10).

Glomus tumor is extremely painful, solitary subcutaneous mass that may involve the subungual region (7). Histologically there are sheets or nests of uniform, round cells with eosinophilic cytoplasm and hyperchromatic round to oval nuclei.

Superficial angiomyxoma may share same histological features with SAFM by presence of spindle-shaped and stellate cells embedded in a vascular, basophilic matrix. However in contrast to SAFM, angiomyxomas are dermal and subcutaneous nodules, which may contain epithelial component, and affect any body part, especially head, neck and trunk. Again, tumor cells are positive for vimentin (10).

Acral fibrokeratomas are exophytic, solitary lesions. In contrast to SAFM, they are mainly composed of paucicellular vertically interwoven collagen bundles with a collarlette (1, 2).

Sclerosing perineuroma is a variant of perineuroma that affects fingers and palms of

young adults. It is composed of dense collagen with small, epithelioid and spindle-shaped cells arranged in cord, trabecular and onion-skin pattern (1, 2, 4, 10). This is quite distinct from SAFM.

Cutaneous myxoma, may be solitary, involves the digits. These tumors have a mucinous matrix with fibroblasts and prominent capillaries, but in contrast to SAFMs, these lesions may contain epithelial component (10).

Myxoid fibrous histiocytomas contain spindle cells in the storiform pattern and sclerotic collagen at the periphery of the lesion. The myxoid area is not abundant as it is in SAFMs and is positive for factor XIIIa antigen, but not for CD34 (11).

Low grade myxofibrosarcoma is a slow-growing tumor usually located in the subcutaneous tissues of extremities. This tumor contains small stellate cells with pleomorphic nuclei around the blood vessels and pseudolipoblasts (9).

There are no reported cases of FNAC diagnosis of SAFM in the literature. The classic location of this tumor, along with aspiration of a cellular population of spindle cells admixed with myxoid background elements enabled a correct diagnosis. Immunocytochemistry may prove useful in the future.

The pathogenesis of SAFM is also much debated. Recent studies proposed multipotential dermal stem cells or mesenchymal cells in the nail unit as the probable cell of origin based upon expression of nestin (marker of multipotent stem cells) or CD-10 (co-expressed by mesenchymal cells in the nail unit) by some tumors (12,13).

SAFM may recur or persists if inadequately excised (1). Recurrence rate may be as high as 22% (14). Pre-operative correct diagnosis will help to plan for complete surgical removal. So, although rare, appropriate care must be taken for cytodiagnosis of these rare neoplasms to reduce future complications.

Acknowledgements

The authors declare that there is no conflict of interest.

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