Cytological diagnosis of superficial acral fibromyxoma: A case report

ABSTRACT
Superficial acral fibromyxoma (SAF) is a rare, distinctive benign soft tissue lesion that often involves the fingers and toes, with the great toe being the most frequently affected site. We report a case of SAF diagnosed by fine needle aspiration cytology and confirmed by histopathology. The pre-operative cytological diagnosis will help the surgeon to plan for a wider excision that prevents recurrence.

Key words: Fine needle aspiration cytology; great toe; soft tissue; superficial acral fibromyxoma

Introduction
Superficial acral fibromyxoma (SAF) is a rare, distinctive benign soft tissue tumor with a predilection for the hands and feet. It often involves the fingers and toes, with the big toe being most frequently affected. To date, there is only one case that has been diagnosed on cytological features. We herein report a case of SAF primarily diagnosed by fine needle aspiration cytology (FNAC) and confirmed by histopathological study.

Case Report
A 27-year-old healthy female presented with an asymptomatic, globular swelling over the right great toe since 2 years. Examination revealed a 2 cm × 1 cm × 1 cm hemispherical firm, non-tender nodule in the subungual region with no involvement or deformity of the nail plate [Figure 1].

Fine needle aspiration (FNA) yielded mucoid material, which showed benign plump oval to spindle cells arranged in groups, sheets and cohesive clusters in a background of myxoid material with interspersed thin-walled capillaries. The cells had indistinct cytoplasm and blunt oval to spindle-shaped nuclei [Figure 2]. With these, a diagnosis of SAF was suggested and an en block excision biopsy was performed and subjected to histopathological examination.

Grossly, the specimen consisted of a skin-covered tissue, which on cut-section revealed a well-circumscribed...
grey-white lesion in the subcutis. On microscopy, there was a hyperkeratotic epidermis and a dermal tumor composed of spindle and stellate cells with random, loose storiform and fascicular patterns that were embedded in abundant myxocollagenous stroma that showed Alcian blue positivity [Figure 3]. The cells showed no pleomorphism, mitotic figures or atypia. These features confirmed the diagnosis of SAF.

**Discussion**

The term SAF has been coined recently in 2001 by Fetsche et al.\(^1,3\) It represents a fibrohistiocytic tumor with less than 170 described cases to date. It is not listed in the current World Health Organization’s pathology and genetics of skin tumors, but was listed in the Armed Forces Institute of Pathology’s non-melanocytic tumors of the skin (Atlas of Tumor Pathology).\(^4\)

It presents as a slow-growing tender mass in adult males.\(^4,5\) Most cases occur in the subungual or periungual regions, with rare cases occurring on the ventral surface of the digit.\(^2\) Few cases located close to the nail caused deformity of the nail and affected the underlying bone, but there was no evidence of systemic involvement.\(^6\) Majority of the patients were males, with a male to female ratio of 1.3:1, and they belonged to an age range of 14-91 years with a mean of 48.2 years and a median of 49.0 years.\(^4,6\) As in the present case the tumor presented as a painless solitary mass with an average size of 17.8 mm, few cases that presented with pain were associated with history of trauma.\(^6\) Roentgenographic examination in most cases showed no bony alterations, while exceptional cases showed marginal erosion of distal phalanx or depression of the underlying bone.\(^4,7\)

FNA yielded mucoid material and showed loose clusters of benign spindle cells in a myxoid background.\(^3\) The lesions with no involvement of the underlying nail plate or bone when excised completely revealed a dome-shaped, well-circumscribed tumor in the dermis as seen in the present case.\(^7\)

SAF are generally unencapsulated, but, in the present case, the tumor displayed a vague lobular pattern that compressed the surrounding tissue giving a partially pseudocapsulated appearance.\(^6\) The tumor was composed of spindle- to stellate-shaped cells arranged in a vague storiform pattern embedded in a myxoid matrix.\(^3,4,6\) Often, there is accentuated vasculature and increased numbers of mast cells.\(^4,6\) Multinucleated stromal cells may be present, but nuclear atypia and mitotic figures are rare.\(^4\) Previous studies have shown that the myxoid matrix is highlighted by the Alcian blue (pH 2.5) stain.\(^4,6\)

Characteristically, tumor cells show positivity for vimentin, CD-34, CD-99, EMA and CD10 and are negative for S-100 protein.\(^1,3,4,6\) Expression of nestin (a marker of multipotent stem cells) or CD-10 (co-expressed by mesenchymal cells in the nail unit) points to the cell of origin as being either the multipotent dermal stem cells or the mesenchymal cells in the nail unit.\(^8,9\) In an ultrastructural study by Pasquinelli et al.,\(^10\) the tumor cells were found to be composed of cytoplasmic intermediate filaments and numerous cisternae of rough endoplasmic reticulum, confirming the fibroblastic nature of the tumor cells.

Malignant behavior or metastasis has not been reported, but the tumor may persist or recur if inadequately excised.\(^4\) It is observed that all recurrent tumors have positive margins on initial biopsy or on subsequent excision. A pre-operative correct diagnosis of SAF by FNA cytology is thus essential.
to plan a complete surgical excision that avoids further recurrence. The differential diagnosis for SAF includes myxoid neurofibroma, fibroma of tendon sheath, glomus tumor, superficial angiomyxoma, acral fibrokeratoma, sclerosing perineuroma, cutaneous myxoma, myxoid fibrous histiocytoma and dermatofibrosarcoma protuberans.

Myxoid neurofibroma often has a neural appearance, with no increase in vascularity, and has a characteristic S-100 positivity. Fibroma of the tendon sheath has an attachment to the tendon sheath and has only sparse stellate cells in a fibrocollagenous matrix. The glomus tumor is a very painful dermal nodule composed of sheets or nests of uniform round cells with immunoreactivity for vimentin and smooth muscle actin. Superficial angiomyxomas are dermal or subcutaneous nodules that can affect any part of the body, especially the head, neck and trunk. These tumors, in addition to having spindle-shaped and stellate cells in a basophilic matrix, have an epithelial component. Acral fibrokeratoma are exophytic, solitary lesions with a hyperkeratotic epidermis and a core of thick, vertically oriented collagen bundles. The tumor is paucicellular and EMA negative. Sclerosing perineuroma is composed of dense collagen with small, epitheloid and spindle-shaped cells arranged in cords, trabecular and onion-skin patterns, which make them distinct from SAF. Cutaneous myxoma has an epithelial component in addition to the fibroblasts and prominent capillaries in a mucinous matrix. Myxoid fibrous histiocytoma contain spindle cells in a storiform pattern that show positivity for factor XIIIa antigen and the myxoid area is not as abundant as in SAF. Dermatofibrosarcoma protuberans is a dermal tumor composed of cells arranged in storiform pattern and is often seen extending into the subcutis.[4,5]

**Conclusion**

SAF is not widely recognized because of its uncommon occurrence. Awareness of this entity is helpful in distinguishing this lesion from other myxoid soft tissue tumors and in proper management of these cases.

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