Case Report

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A rare case of superficial acral fibromyxoma

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ABSTRACT

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

Keywords: SAF

INTRODUCTION

Superficial acral fibromyxoma is a rare soft tissue benign neoplasm that has a predilection to develop on hands and feet. Superficial acral fibromyxoma usually involves the fingers and toes, and big toe is the most frequently affected site. Most cases of superficial acral fibromyxoma tend to occur in the subungal or periungal regions, although two rare cases occurring on the ventral surface of the digit have been described. We report a case of superficial acral fibromyxoma, which developed at the big toe.

CASE REPORT

66 years old male patient, a farmer by occupation, presented with c/o progressively increasing swelling over the right great toe - 2 months duration, h/o serosanguinous discharge from the ulceration over the swelling, pt c/o itching with h/o trauma over the same toe 6 months back. Examination revealed a 3x2 cm firm, non-tender swelling in the subungual region with deformity of the nail. A differential diagnosis of squamous cell carcinoma, mycetoma foot, ungal wart, dermatofibrosarcoma protuberens was given. Edge wedge

biopsy was done which showed sections of hyperkeratotic stratified squamous epithelium with underlying spindle cells in a fibromyxoid stroma confirming the diagnosis of SAF. X-rays of the foot showed no involvement of the bone. Hence pt was proceded to disarticulation of the great toe with plantar flap cover; histopathology of the specimen confirmed the diagnosis. Immunohistochemistry showed the neoplastic cells were positive for positive for CD34, CD10, EMA, Nestin and negative for S100, SMA. Post op was uneventful and patient is under follow up.



Figure 1: Pre-operative image of SAF involving right great toe.



Figure 2: Pre-operative image of SAF of right great toe.

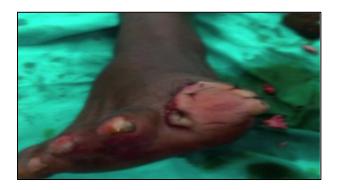


Figure 3: Intra-operative image of plantar flap.



Figure 4: Intra-operative image of plantar flap.



Figure 5: Excised specimen.



Figure 6: Post-operative image.

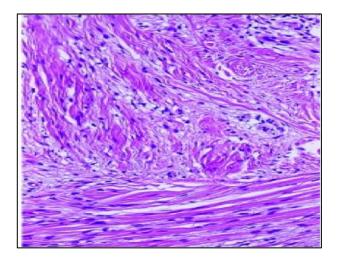


Figure 7: Histology slide showing proliferated spindle and stellate cells with random and fascicular patterns in the myxoid stroma.

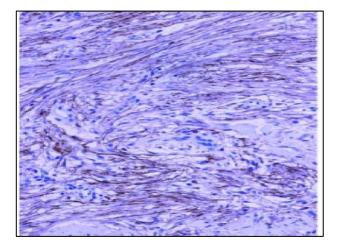


Figure 8: Vimentin positivity.

DISCUSSION

Clinically the important points in superficial acral fibromyxoma are a frequent deformity of the nail plate, a

need for the removal of the nail plate during surgical procedures and a rare deformity of the bone. However there is no evidence systemic involvement

Majority of the patients were males, with a male to female ratio of 1.3:1. As in the present case the tumor usually presents as a painless mass, however few cases that presented with pain were associated with history of trauma, roentgenographic examination in most cases showed no bony alterations, while some cases showed marginal erosion of distal phalanx or depression or scalloping of the underlying bone. 11,12

The histogenesis of superficial acral fibromyxoma is still unclear. Expression of CD-10 has been reported in normal mesenchymal cells in the nail unit.⁷ Recent investigations have revealed that multipotent precursors isolated from the dermis express nestin.^{8,10} Neoplastic cells in this condition are related to mesenchymal cells in the nail unit (onychoblasts) and show dedifferentiation into dermal stem cells.

Characteristically, tumor cells on immunohistochemistry show positivity for vimentin, CD-34, CD-99, EMA and CD10 and are negative for S-100 protein. Expression of nestin which is a marker of multipotent stem cells or CD-10 which is co-expressed by mesenchymal cells in the nail unit points to the cell of origin as either multipotent dermal stem cells or the mesenchymal cells in the nail unit. In an ultra-structural study by Pasquinelli et al which found that the tumor cells were composed of cytoplasmic intermediate filaments and numerous cisternae of rough endoplasmic reticulum, confirming the fibroblastic nature this tumour cells.¹³

Malignant behavior or metastasis has not yet been reported, but recurrence is common if inadequately excised. 12 It is found that all recurrent tumors have positive margins on initial biopsy or on subsequent excision. A preoperative diagnosis of SAF by FNA cytology is thus essential to plan for a complete surgical excision that avoids further recurrence. The differential diagnosis for SAF include myxoid neurofibroma, fibroma of tendon sheath, superficial angiomyxoma, acral fibrokeratoma, glomus tumor, sclerosing perineuroma, myxoid fibrous histiocytoma, cutaneous myxoma, myxoid fibrous histiocytoma and dermatofibrosarcoma protuberans.

Myxoid neurofibromas often have a neural appearance, with no increase in vascularity, and have a characteristic S-100 positivity. Fibromas of the tendon sheath have attachment to the tendon sheath and have only sparse stellate cells in a fibrocollagenous matrix. Glomus tumour is a painful dermal nodule composed of nests of uniform round cells with immunoreactivity for vimentin and smooth muscle actin. Superficial angiomyxomas are dermal or subcutaneous nodules that affect any part of the body, especially the head, neck and trunk. They also have an epithelial component. Acral fibrokeratoma are

exophytic, lesions with a hyperkeratotic epidermis and a core of thick, vertically oriented collagen bundles. The tumor cells are paucicellular and EMA negative. Sclerosing perineuroma is composed of dense collagen with small, epitheloid spindle shaped cells arranged in cords with onion-skin pattern, which make them distinct from SAF. Cutaneous myxomas have fibroblasts and prominent capillaries in a mucinous matrix along with an epithelial component. Myxoid fibrous histiocytoma contain spindle cells in a storiform pattern and have positivity for factor XIIIa antigen and the myxoid area less abundant than in SAF. Dermatofibrosarcoma protuberans is a dermal tumor composed of tumour cells arranged in storiform pattern and often subcutaneous extension is seen. 12,14

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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institutional ethics committee

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