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Case Report

Surgery Section

Superficial Acral Fibromyxoma: A Rare Entity - A Case Report

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ABSTRACT

Superficial acral fibromyxoma (SAF) is a rare lesion initially described by Fetsch as a tumour with histological and immunohistochemical features located at acral sites. It is a benign slow-growing soft tissue lesion occurring in males. Patients generally look for late medical help as it is generally painless. The lesion consists of spindled and stellate-shaped cells in the myxocollagenous stroma with immunohistochemical positivity for CD34, CD99 and vimentin. Treatment is surgical excision with regular follow-up. Here we present a case of a superficial acral fibromyxoma of the left index finger which radiologically showed features of vascular anomaly on MRI. The lesion was surgically excised and histopathology revealed features suggestive of superficial fibromyxoma with positive immunohistochemistry for CD34 and negative for S-100.

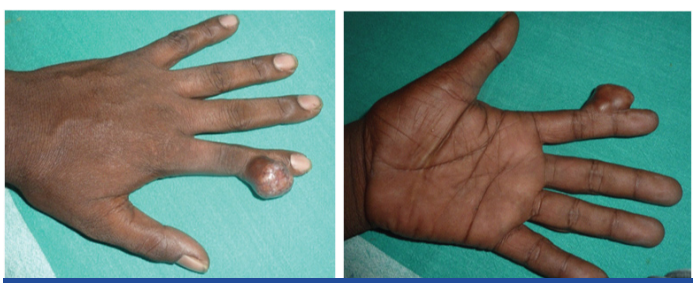
Keywords: Asymptomatic, Benign, Immunohistochemistry, Surgical excision

CASE REPORT

A 31-year-old male came to the Department of Plastic Surgery with a swelling at the tip of the left index finger for about 8 years and pain in the same site for about 15 days. It started as a small pea sized swelling and has gradually progressed to the present size. There is no history of trauma to the left index finger tip. On examination, a tender swelling of approximately 5x3 cm was present along the dorso-radial aspect of left index finger in the region of the middle phalanx, with well-defined margins and a variegated irregular surface. It was firm in consistency and non – fluctuant. Finger movements were normal with no distal neurovascular deficit [Table/Fig-1,2].

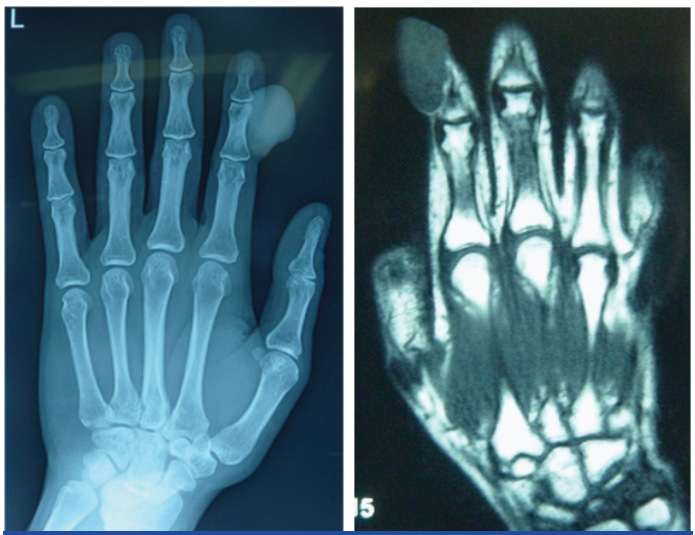
Plain radiograph showed a homogenous soft tissue swelling over the radial aspect of the middle phalanx with no bony involvement [Table/Fig-3]. MRI revealed a complex multiloculated lesion which was hypointense on T1W [Table/Fig-4], corresponding hyperintense on T2W [Table/Fig-5] and intensely bright on Inversion Recovery (IR) sequence, measuring 2.7x2.1x2.5 cm. It was a predominantly cystic lesion in the dorsolateral aspect of left index finger. There were thin hypointense septations seen with thin hypointense focal area of blooming, likely calcification in the posterior part of the lesion. The lesion arose from the skin and subcutaneous plane and abuts the middle phalanx of the 2nd digit. Medially it abuts the joint capsule of the left index distal interphalangeal joint and collateral ligament. A delayed intense enhancement was seen on contrast administration [Table/Fig-6]. The features are likely to be a vascular lesion mostly a cavernous haemangioma.

Surgical excision was planned under regional anaesthesia and tourniquet control. The lesion was excised with a healthy margin of normal tissue leaving behind the intact radial neurovascular bundle.

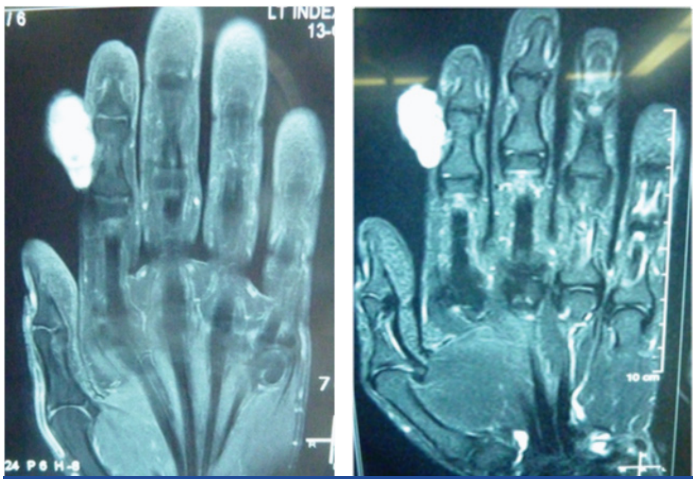


[Table/Fig-1]: Clinical photograph showing the lesion – dorsal view.
[Table/Fig-2]: Clinical photograph showing the lesion – palmar view.

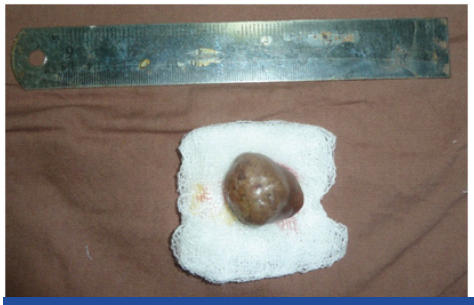
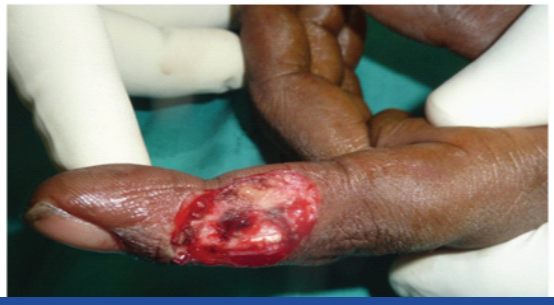
The specimen was sent for histopathological examination [Table/Fig-7]. The resultant defect [Table/Fig-8] was covered with a full thickness skin graft harvested from the cubital fossa [Table/Fig-9].



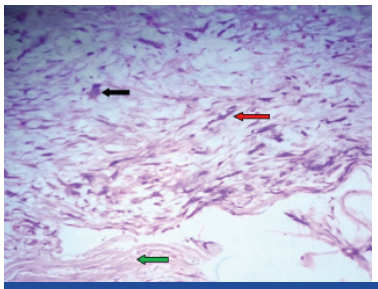
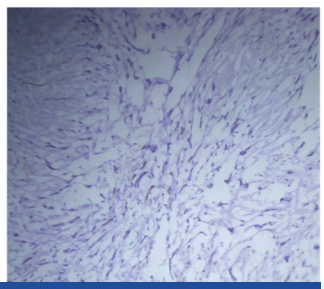
[Table/Fig-3]: Plain radiograph of the left hand showing the lesion in the radial aspect of the left index finger. **[Table/Fig-4]:** T1 weighted image of MRI showing hypointense lesion with fine fibrous septations.



[Table/Fig-5]: T2 weighted image of MRI revealing the with hyperintense lesion.
[Table/Fig-6]: MRI showing intense enhancement on contrast administration.


[Table/Fig-7]: Specimen following excision. **[Table/Fig-8]:** Defect following excision. **[Table/Fig-9]:** Photograph showing the defect covered with a full thickness skin graft.

[Table/Fig-10]: Proliferation of spindle (red arrow) and stellate-shaped (black arrow) cells in a myxoid stroma (green arrow) (H&E 40X).
[Table/Fig-11]: Immunohistochemical staining showing positivity for CD 34 (H&E x400 magnification).

The patient was discharged on 5th postoperative day; graft taken was 100%, following which the patient was lost to follow-up.

On histopathology, macroscopy revealed skin covered soft tissue measuring 2.5x3 cm with cut surface showing multiple cystic areas and glistening white material. Microscopy showed hyperkeratotic epidermis with lesion beneath composed of spindle and stellate shaped cells, seen in sheets, fascicles and vague whirling pattern in an abundant myxomatous stromal background [Table/Fig-10]. There were intervening fibrous septae, occasional mast cells and multinucleated stromal cells, features suggestive of superficial fibromyxoma – suggested IHC for conformation. Immunohistochemical staining was positive for CD34 [Table/Fig-11] and negative for S-100.

DISCUSSION

Superficial acral fibromyxoma (SAF) is a rare, benign fibromyxoid tumour commonly located in the hands and feet, initially described by Fetsch et al., [1,2]. It generally presents as a slow-growing, nodular mass commonly found in the toes than the fingers, mostly involving the subungual or periungual region, the other involved sites being the heel, palm and ankle [1,3-6]. The lesion affects males twice as often as females, generally in the fifth to sixth decade. No specific cause has been attributed but history of trauma is rare and has been reported [7]. Due to the asymptomatic and painless nature of these lesions, there is a delay in presentation and diagnosis. Differential diagnosis includes acquired digital fibrokeratoma, periungual and subungual fibroma, superficial angiomyxoma, dermal mucinosis, myxoid neurofibroma, sclerosing perineuroma, myxoid dermatofibrosarcoma protuberans, low grade fibromyxoid sarcoma, fibroma of tendon sheath, glomus tumour, giant cell tumour, fibrous histiocytoma, cutaneous myxoma and inflammatory myxohyaline tumour of distal extremities [8]. Acquired digital fibrokeratomas are solitary tumours of fingers and toes positive for CD34 but negative for Epithelial Membrane Antigen (EMA). Superficial angiomyxoma are located on the head, neck or trunk and are CD 34 and S-100 positive. Myxoid neurofibromas are solitary lesions of the hands and feet but are S-100 positive. Sclerosing perineuromas are acral benign fibrous tumours positive for EMA and negative for S-100 and CD 34. Myxoid dermatofibrosarcoma protuberans are rare aggressive mesenchymal tumours more centrally located which are CD 34 positive and S-100 negative. Low grade fibromyxoid sarcoma is rare in acral regions. Inflammatory myxohyaline tumour has characteristic virocyte or Reed-Sternberg cells [8].

Macroscopically, the tumour is firm, nodular or lobulated mass with a grayish-white cut surface, the consistency being solid to gelatinous. The tumour usually involves the dermis and the subcutaneous tissue. Fascial extension is rare, but involvement of periosteum causes erosion of the underlying bone [9]. Microscopically, it is a well-circumscribed dermal or subcutaneous tumour with increased vascularization and without a capsule [2,9-11]. The lesion consists of spindle and stellate cells arranged in a loose storiform pattern or in fascicles embedded in a myxoid, collagenous, or myxocollagenous stroma [2]. Mast cells often are prominent, with an increased number of ecstatic blood vessels. Inflammatory infiltrates and multinucleated giant cells typically are not seen [12]. On immunohistochemistry, tumour cells typically express CD34, CD99, vimentin and EMA and are negative for S-100 protein, smooth muscle actin, desmin, cytokeratin, glial fibrillary acidic protein, apolipoprotein D, and Human Melanoma Black 45 (HMB-45) [1-3,5,9,10]. Expression of CD10 was reported in a small case series study of 4 cases by Tardı´o et al., [13].

Management of these tumours involves surgical excision to rule out malignancy and prevent recurrence. There have been no reported cases of malignant change in the literature [2]. Regular follow-up after excision is warranted as the recurrence rate reported is as high as 24% mostly due to incomplete excision [3,5].

CONCLUSION

SAF is a rare benign and indolent lesion recently described by pathologists and surgeons should be aware to include it in the differential diagnosis of fibromyxoid tumours of acral sites. Early diagnosis, complete resection and strict follow-up are the key to prevent recurrence.

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