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"SUPERFICIAL ACRAL FIBROMYXOMA - AN UNUSUAL VISITOR TO A SURGEON"

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ABSTRACT

Superficial acral fibromyxoma is a very rare benign slow growing myxoid neoplasm identified in recent years. It has a predilection for ungual region of fingers and toes in middle aged adults, presenting as a painless polypoid slow growing nodule or mass involving distal part of finger or toes. It i s a well circumscribed, non-encapsulated dermal neoplasm composed of spindle shaped cells embedded in a myxoid or collageous matrix without significant nuclear pleomorphism. Tumour cells are immunoreactive for CD34 and CD99 and vimentin but variable sensitivity to epithelial membrane antigen (EMA). Treatment consists of wide excision, although malignant transformation has not been documented. Regular follow up is essential as the recurrence rate is reported to be around 24%.

KEY WORDS

Benign, Periungual and subungual, Soft tissue tumour, Fibromyxoma, Excision

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INTRODUCTION

Superficial Acral Fibromyxoma (SAF) is a very rare slow growing soft tissue tumour, first described in 2001 by Fetsch et al[1]. It involves the periungual and subungual regions of fingers and toes, occuring mainly in middle aged males[3]. Histology shows a dermal lesion with spindle shaped fibroblasts in storiform or fasciculated pattern in myxocollagenous stroma[2]. Immunohistochemistry usually is positive for CD34 and CD99 with variable positivity for epithelial membrane antigen[2]. Wide excision with tumour free margin is the treatment of choice as the recurrence rate is advocated as 24%. Very few cases have been reported worldwide and only a couple in Indian population[3,6].

CASE REPORT

A 65 year old female patient presented with swelling over right middle finger, gradually progressive over last 15 years. There was no history of pain and trauma. Local examination revealed a globular swelling 3x4 cm in dimensions, involving distal phalanx of right middle finger circumferentially. The surface was irregular and consistency firm along with thinning and broadening of nail.

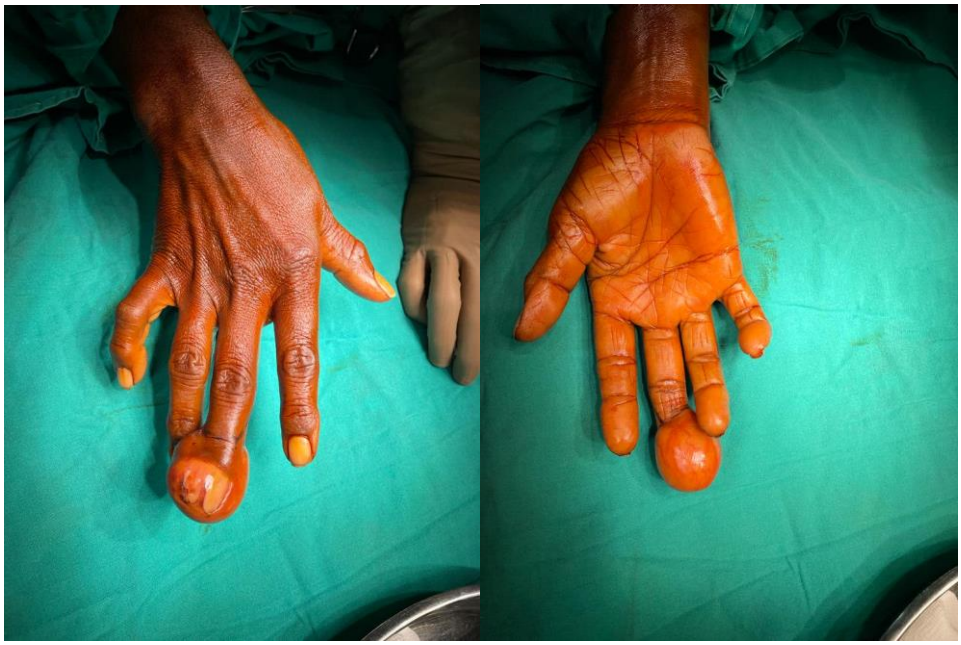

Fig. 1 : Clinical photographs of a case of Fibromyxoma involving Right middle finger.

Investigations were within normal limits. X-Ray of right middle finger revealed slight periosteal erosion of terminal phalanx along with the variegated appearance of Fibromyxoma. FNAC was suggestive of nonspecific benign fibromatous lesion. Immunohistochemistry revealed positivity for CD34 and CD99.

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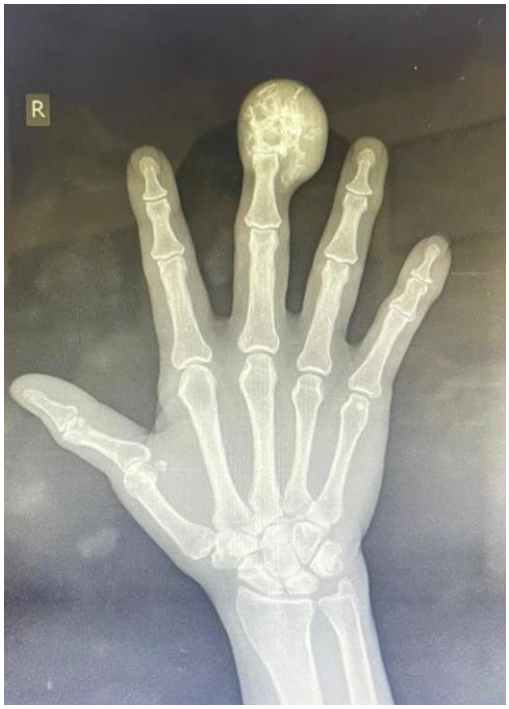

Fig. 2 : Plain X-Ray film of right hand showing Fibromyxoma involving the tip of the right middle finger.

Wide excision of the lesion was performed with amputation of distal phalanx including terminal part of middle phalanx.

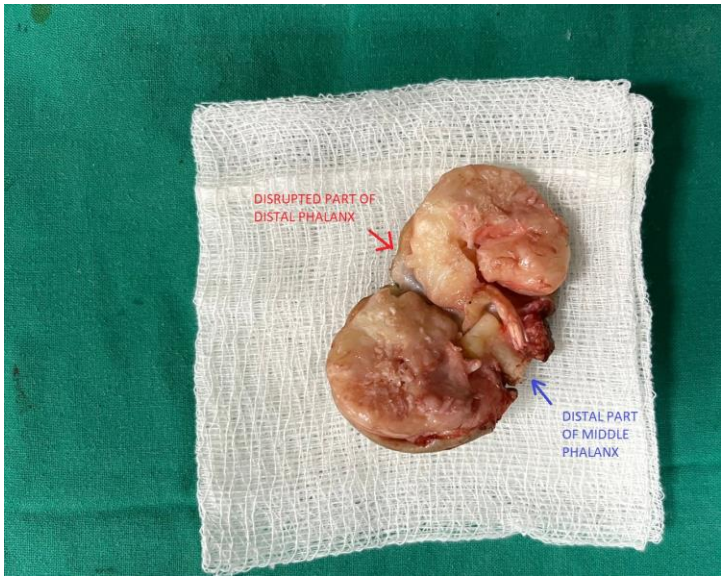

Fig.3 : Excised specimen of Fibromyxoma involving the terminal part of right middle finger.

The histopathology revealed superficial acral fibromyxoma. Postoperative course of the patient was uneventful.

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DISCUSSION

Superficial acral fibromyxoma (SAF) is a very rare, benign, myxoid tumour of soft tissues affecting periungual and subungual regions of fingers and toes[3]. It is also called Digital Fibromyxoma[5]. It was presented as a distinct clinicopathological entity in 2001 by Fetsch et al[1], who also did immunohistological analysis of 37 cases. Since then not many cases have been reported worldwide and only a few in Indian population[3].

SAF usually presents as a painless mass of slow growth affecting mainly males in 5th decade of life[2], although in our case the patient was a female which is very rare. History of associated trauma is very rare[1] and most tumours affect acral region of hands and feet, growing very slowly and mostly being asymptomatic[7,8].

It is usually a polypoid dome shaped well circumscribed tumour affecting distal part of fingers or toes, but can affect heel or ankle[2]. It is usually encapsulated and located in dermis and subcutaneous tissues[5]. External surface can be nodular or irregular while cut section shows gelatinous grey white appearance[4]. Fine Needle Aspiration Cytology shows loose cluster of spindle cells in myxoid material[6]. Histological examination shows benign neoplasm composed of spindle and stellate shaped fibrobastic cells arranged in loose storiform or fascicular pattern, embedded in loose myxoid or collagen stroma[3,6]. It also often shows marked microvascularisation and mast cells, but mitotic figures are infrequent[2].

Radiological examination may show underlying bony erosion and scalloping due to mass effect of the tumour[6]. Ultrasound examination can add important information about mass size, location and content, complemented by doppler examination which may enhance vascular pattern. Magnetic Resonance Imaging demonstrates homogeneous hyperintensity in T2 weighted images and increased contrast to normal tissue[2,10].

Immunohistochemistry shows tumour cells staining positive for CD99, CD34 and vimentin, while immunoreactivity to Epithelial Membrane Antigen (EMA) is variable[4].

Differential diagnosis include CD34 immunopositive neoplasm such as Dermatofibrosarcoma Protuberans, Superficial Angiomyxoma and Myxoid Neurofibroma and CD34 immunonegative tumours such as Sclerosing Perineuroma, Fibrous Histiocytoma, Glomus Tumour and Giant Cell Tumour of tendon sheath[4].

Dermatofibrosarcoma Protuberans is a dermal tumour commonly extending into subcutis (as opposed to SAF) composed of spindle cells arranged in storiform pattern interspersed with extensive myxoid areas, being positive for CD34 and Vimentin but negative for EMA[4,10]. Myxoid Neurofibroma has a neural appearance but no increase in vascularity, characterised by cells that are positive for S-100 mixed with CD34 positive fibroblastic cells which differs from SAF which is S-100 negative and has concentrated microvasculature[2].

Treatment of SAF involves wide surgical excision to prevent malignancy and recurrence[2], although malignant transformation has not been documented[4]. In our case amputation was performed at the level of middle phalanx for a complete wide excision of the lesion.

Regular follow up after excision should be done meticulously as the recurrence rate is reported to be 24%[8] mainly due to incomplete excision. Thus, although very rare, SAF may be encountered quite unexpectedly in surgical practice. Hence, SAF should be always included in the differential diagnosis of any slow growing mass or nodule affecting periungual or subungual regions of fingers or toes[2,9].

CONCLUSION

Superficial Acral Fibromyxoma is a slow growing myxoid soft tissue tumour with a predilection of acral region and should always be considered when a patient presents with a periungual or subungual slow growing nodule or mass affecting fingers or toes. It is a dermal or subcutaneous tumour composed of spindle or stellate cells within myxoid or collagenous stroma with prominent blood vessels or mast cells. Treatment involves wide surgical excision and regular follow up for early detection of recurrence, which is reported to be around 24%.

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