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Superficial Acral Fibromyxoma: A case report

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ABSTRACT

INTRODUCTION: Superficial Acral Fibromyxoma is a benign and rare tumor of the soft tissues, also it called digital fibromyxoma. It is a painful, slow growing solitary mass that is frequently observed in the fingers, toes and nail beds. Local recurrence is often associated with a positive margin.**PRESENTATION OF CASE:** A 54-year-old male patient had admitted to local state hospital with complaint of a two-lobed mass lesion in the 5th finger of the right hand. The lesion was closely adjacent to the 5th finger flexor tendon. The specimen of the patient undergoing excisional biopsy was sent to our pathology department.**CONCLUSION:** Superficial Acral Fibromyxoma is one of the rare benign myxoid neoplasms. Its treatment is surgical excision.

We reported the case for its rarity and to keep in mind in the acral region neoplasms.

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1. Introduction

Superficial Acral Fibromyxoma (SAF) was first described by Fetsch in 2001 as a localized tumor in acral extremities. SAF is a rare clinical occurrence that generally occurs in the fingers, toes and nail beds [1,2]. It is a benign neoplasm that usually grows slowly and is seen in men [1,3]. Since they are generally painless, medical application is late [1,4]. Trauma history has been described in several cases [4].

This lesion is composed of spindle and star-shaped cells that are positive for CD34, CD99 and Vimentin immunohistochemically in myxocollagenous stroma [1,5]. Tumor cells are arranged as loose storiform and focal fascicular pattern. In SAF, nuclear pleomorphism is mild or moderate, mitotic activity is rare. Sometimes multinuclear cells can be seen [1,6]. Its treatment is surgical excision and requires regular follow-up [1,3]. This work has been reported in line with the SCARE criteria [7].

2. Case report

A 54-year-old male patient had admitted to local state hospital with complaint of a two-lobed mass lesion in the 5th finger of the right hand (Figs. 1, 2).

Abbreviations: SAF, Superficial Acral Fibromyxoma.

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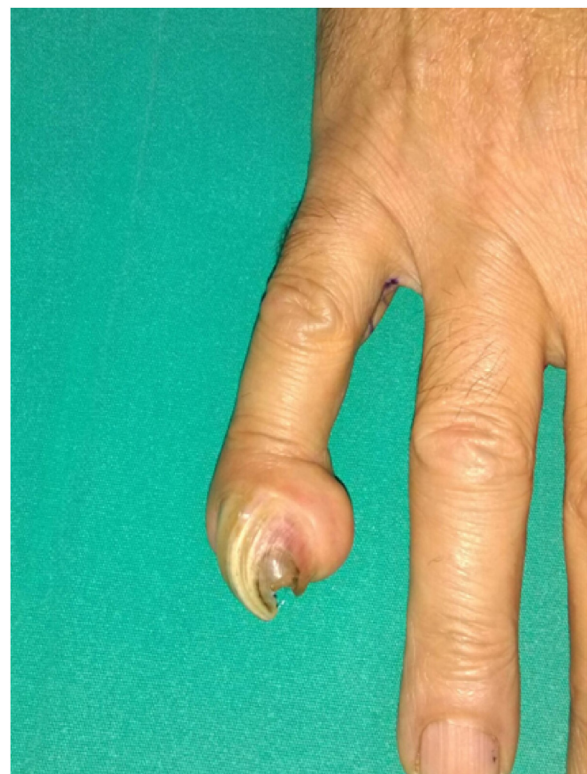


Fig. 1. Clinical photograph showing the 5. finger tip lesion on the right hand with dorsal view.

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Fig. 2. Clinical photograph showing the 5. finger tip lesion on the right hand with palmar view.

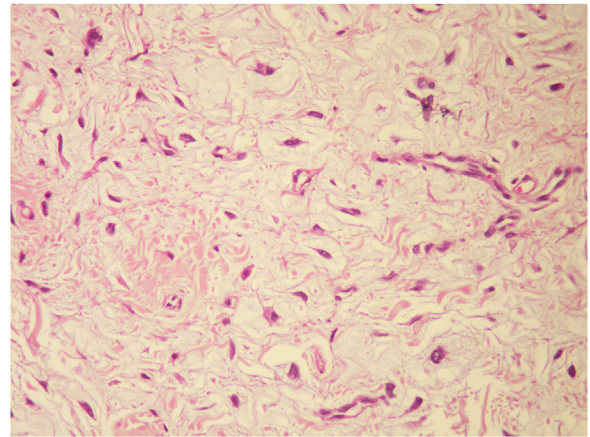


Fig. 4. Spindle-shaped fibroblast-like tumor cells embedded in a myxoid stroma (H&E, ×200).

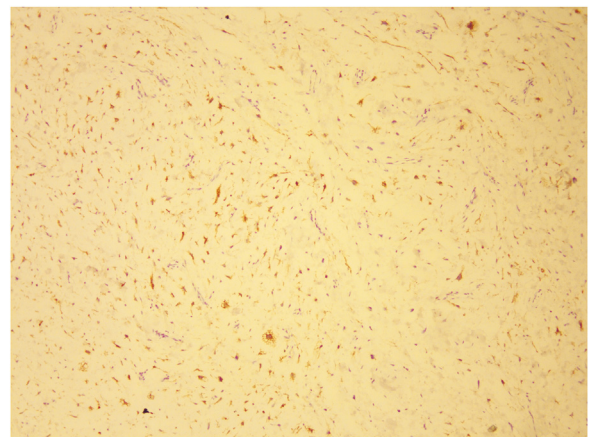


Fig. 5. In neoplastic cells, nuclear positivity for EMA (EMA, ×100).

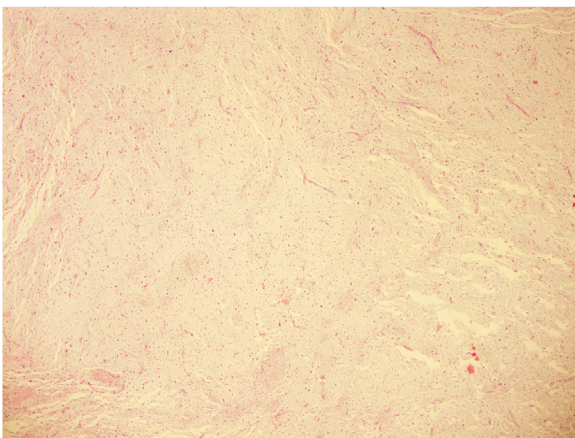


Fig. 3. Tumor cells within a myxoid stroma with prominent vessels (H&E, ×40).

With superficial tissue ultrasonographic examination, solid lesion with dimensions of 24 × 11 mm was observed. The lesion was described with its a hypo-isoechoic heterogeneous character, sharply limited in subcutaneous location. The lesion was closely adjacent to the 5th finger flexor tendon. The specimen of the patient undergoing excisional biopsy was sent to our pathology department. The excised soft tissue piece was grey white in colour, measuring 2.5 × 1.6 × 0.9 cm.

Histopathologically, there was moderate cellularity in the lesion and the tumor was composed of stellate and spindle-shaped fibroblast-like tumor cells embedded in a myxoid matrix (Figs. 3, 4).

With histochemical study, Alcian Blue showed positive staining in mucinous areas and Trichrome showed positive staining in fibrous tissue.

With immunohistochemical study, EMA showed positive staining (Fig. 5), S100 and GFAP negative staining in neoplasm. Ki 67 proliferation index was observed as 1%.

The case has been reported as Superficial Acral Fibromyxoma with its histopathological and immunohistochemical features.

3. Discussion

SAF was first described by Fetsch in 2001 [1,2]. In 37 cases of Fetsch, the nuclear atypia was mild and moderate, atypia was evident only in 3 cases. In our case, there was no nuclear atypia. In Fetsch's study, immunohistochemically, CD34 (21 of 23 cases), EMA (18 of 25 cases), CD99 (11 of 13 cases) were positive, while S100 showed poor staining in only one of 23 cases. In our case, EMA was positive and S100 was negative. SAF typically occurs in the middle-aged adult men (mean age: 46). There is male domination in cases (M/F: 2/1) [1]. Age and gender were compatible with our case. The history of trauma has been described only in a few cases [4]. There was no trauma history in our case.

SAF is usually unencapsulated and well-circumscribed. Family history is generally not expected [1]. This neoplasm is located in the dermis and subcutaneous tissue [1,8]. It is characterized by spindle-shaped cells in the loose storiform or fascicular pattern [1]. In differential diagnosis, low grade myxofibrosarcoma, angiomixoma, neurofibroma, dermatofibroma, dermatofibrosarcoma protuberans should be considered. Immunohistochemical methods play an important role in differential diagnosis [1,9]. Low grade myxofibrosarcoma tends to be in the extremities, may be CD34 positive. It

shows a pronounced nuclear atypia and EMA is negative [1,5,8]. In our case, EMA was positive. Superficial angiomyxoma; mostly seen on the head-neck and trunk, it is diffuse and myxoid and contains neutrophils. In the neurofibroma, there is no vascular image and S100 is positive. In our case, S100 was negative. In the dermatofibroma, the storiform growth pattern is well-circumscribed, Factor 13a and actin is positive, CD34 is negative or focal positive. Dermatofibrosarcoma protuberans, like SAF, are positive with CD34 and CD99, but are rarely superficial and acral localized, Dermatofibrosarcoma protuberans form larger masses, subcutaneous adipose tissue infiltration is typical.

4. Conclusion

Superficial Acral Fibromyxoma is one of the rare benign myxoid neoplasms identified in recent years [1,3,5]. Benign soft tissue tumors are frequently observed clinically. When soft tissue neoplasms are observed on the toes or fingers, Superficial Acral Fibromyxoma, one of the rare myxoid neoplasms should be considered in the differential diagnosis. Its treatment is surgical excision. Follow-up is recommended due to the risk of recurrence [1,3].

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethics approval

The study is exempt from ethical approval in your institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Mürüvvet AKÇAY ÇELİK: Collected the data, design the manuscript and wrote the paper.

Havva ERDEM: Design the manuscript.
Nurten TURHAN HAKTANIR: Collected the data.
The authors read and approved the final manuscript.

Registration of research studies

N/A.

Guarantor

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