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Clinical and Histopathological Mismatch: A Case Report of Acral Fibromyxoma

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Abstract

Background: Acral fibromyxoma (AFM) is a rare benign soft tissue tumor which is described as fibromatous and myxoid tumor of skin and soft tissue. Case details: A 40 year old male presented to Dematology outpatient department with swelling over the wrist for one year duration.The swelling was associated with mild pain and it gradually increased in size to reach the present size. Cutaneous examination revealed a 2x2cm mobile,cystic to firm, non-tender swelling over dorsum of right wrist. Based on its location and clinical feature,it was provisionally diagnosed to be ganglion cyst and excision biopsy was done. Histology showed stellate shaped cells in myxoid background with round to oval nuclei having small inconspicious nucleolus.

Conclusion: Acral fibromyxoma presents a distinct histopathology including a myxoid stroma and spindle-shaped cells, which are essential for accurate diagnosis and management.

Keywords: Acral fibromyxoma; fibromatous; myxoid tumor; Dermatopathology

Introduction

Acral fibromyxoma(AFM) is a rare soft tissue tumor which is a fibromatous and myxoid tumor typically arising in the acral regions likes digits,wrist joint but rare locations can be ankles,legs with a male predeliction. Its exact incidence is not well documented but it is thought to be very uncommon compared to other soft tissue tumors [1]. The tumor is more frequently observed in adults, but rare cases have been reported in reported in children and older adults. It is characterized by a unique histopathology and molecular genetics that can aids in its diagnosis and differentiation from other fibromatous neoplasms of skin and soft tissue but still molecular genetics is not well studied [2].The tumor is characterized by a myxoid stroma and proliferation of spindle-shaped cells. Recent immunohistochemical studies have shown that these tumors typically express markers such as CD34, which aids in distinguishing them from other soft tissue tumors [3-5]. Local recurrence has been found in 22-24% of cases but malignant transformation has not been reported. AFM generally has favourable prognosis with benign course [1, 4]. A thorough literature study revealed that few cases has been reported with this clinical and histopathology mismatch. We present this case because of its classic histopathological findings which help us for final diagnosis.

Case Report

A 40 year old male presented to dermatology outpatient department with swelling over the wrist for one year duration. The swelling was associated with mild pain and it gradually increased in size to reach the present size. Cutaneous examination revealed a 2x2cm mobile, cystic to firm, non-tender swelling over dorsum of right wrist (Figure 1). Based on its location and clinical feature, it was provisionally diagnosed to be ganglion cyst and excision biopsy was done. Histology showed stellate shaped cells in myxoid background with round to oval nuclei having small inconspicious nucleolus (Figure 2, 3, 4). Final diagnosis was given as AFM after ruling out other differentials including myxoid neuromas, superficial angiomyxoma, DFSP, glomus tumor. Patient was on follow up but there had been no recurrence for past one year.

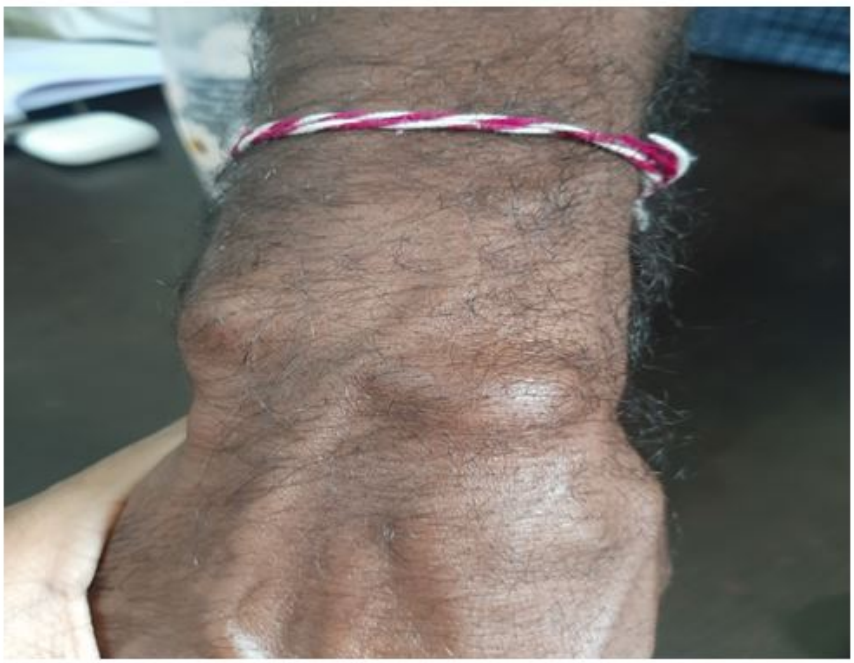

Figure 1: Showed 2x2cm lesion on dorsal surface of the wrist

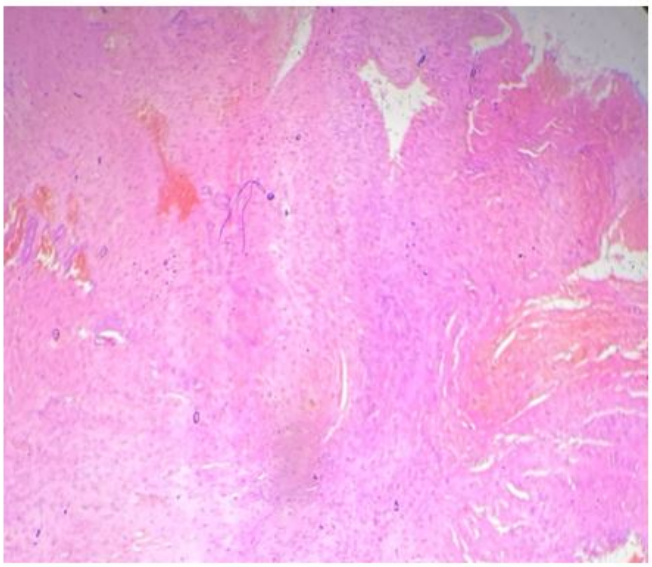

Figure 2: Showed benign looking cells of AFM (H & E stain, 4x)

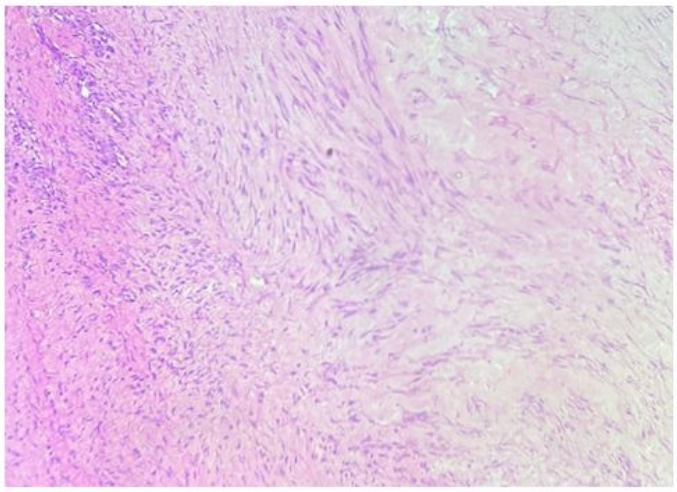

Figure 3: Showed stellate shaped cells in a mxyoid background (H & E stain, 10x)

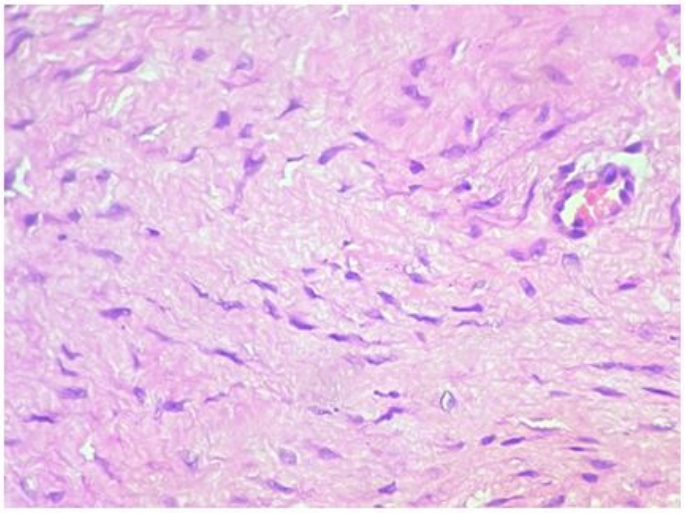

Figure 4: Showed stellate shaped cells without atypia (H & E stain, 40 x)

Discussion

AFM often presents as a painless, firm, solitary sometimes pedunculated mass. The most common locations for AFM are distal parts of limbs like fingers and toes. Less common sites are palms, soles, wrist, hand and thigh. The exact etiology and predisposing factor is not well established [1]. Recent studies emphasize the importance of imaging techniques like MRI for better characterization of the tumor, although definitive diagnosis usually requires histopathological examination and IHC study [3, 5]. The tumor follows a benign course with recurrence rate of 22-24% and no case has been reported showing malignant change [1]. Histologically, AFM is composed of a myxoid stroma with a proliferation of spindle-shaped cells. The tumor typically exhibits a well-circumscribed, non-encapsulated growth pattern. The myxoid matrix is the hallmark feature and is characterized by a loose, gelatinous consistency that provides a striking contrast to the densely cellular components [3]. The spindle cells are often arranged in a storiform pattern and are interspersed with small blood vessels. The nuclei of these spindle cells are usually bland, with minimal pleomorphism. This feature helps to differentiate AFM from more aggressive soft tissue tumors, such as sarcomas [5]. Immunohistochemical staining plays a crucial role in the diagnosis of AFM. The tumor cells typically express markers such as CD34, which is indicative of their fibroblastic lineage. Additionally, they may also show positivity for smooth muscle actin (SMA) in some cases, reflecting myofibroblastic differentiation. However, they are usually negative for S100 protein, desmin, and keratin, which helps rule out other potential diagnoses such as dermatofibroma or nerve sheath tumors [2, 5].

The histopathological features of acral fibromyxoma necessitate careful differentiation from other soft tissue tumors, including dermatofibromas, myxofibrosarcomas, and other fibromyxoid lesions. The myxoid stroma and the specific cellular arrangement are key aspects that pathologists consider in making an accurate diagnosis. The absence of significant nuclear atypia and the presence of the characteristic myxoid matrix are crucial points that help to distinguish AFM from more aggressive tumors [2].

Recent studies have identified genetic mutations, particularly in the loss of RB1 gene, associated with acral fibromyxoma.While genetic testing is not routinely performed in clinical settings, it may eventually offer additional diagnostic and therapeutic avenues [2, 3]. Understanding the histopathological aspects of acral fibromyxoma is essential for effective management. Complete surgical excision is the primary treatment modality, and the prognosis is generally favorable. But patient should be kept under follow up because few cases show recurrence.However, careful histopathological evaluation is critical, as incomplete excision can lead to recurrence, reported in up to 25% of cases [2].

Conclusion

Acral fibromyxoma presents distinct histopathological characteristics, including a myxoid stroma and spindle-shaped cells, which are essential for accurate diagnosis and management.IHC like CD 34 is not diagnostic or unique to AFM but requires combination of clinical,histomorphology. An understanding of its genetic may improves diagnostic accuracy and may provide potential therapeutic strategies but still research has to be done on the genetic and definitive diagnostic criteria for AFM.

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