Solitary Firm Papule Over Periungual Area

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Case History

An adult female presented with complain of single-well defined pink to erythematous firm papule of size 0.5 × 0.5 cm over proximal nail fold of left index finger from 1 year [Figure 1]. There was no history of prior trauma, bleeding or ulceration. On palpation, it was firm and non-tender with no attachment to underlying tissue. Roentogram of left hand did not show any underlying bony abnormality. Lesion was completely excised under ring block anesthesia with clear resection margins. Excised growth was sent for histopathological examination which showed hyperkeratosis and parakeratosis in epidermis [Figure 2]. Dermis showed proliferation of spindle to stellate cells in a myxoid matrix with prominent vascular pattern [Figure 3]. No mitotic figures seen. On immunohistochemistry tumor cells are CD99+ and CD34 [Figure 4 and 5], other stains such as vimentin and S-100 could not be performed.

What is your diagnosis?

Answer:

Superficial acral fibromyxoma

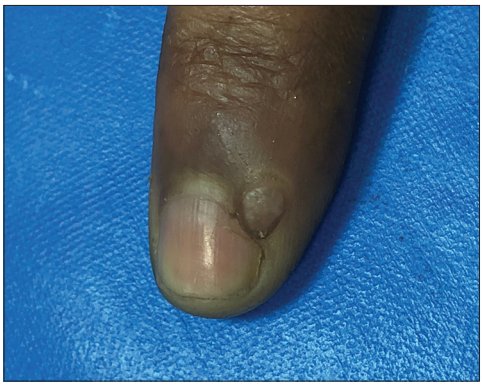

Figure 1: A solitary skin colored firm papule over proximal nail fold area of the index finger

Discussion

Superficial acral fibromyxoma is a rare soft tissue tumor, which was first described in year 2001 by Fetsch *et al*.[1] First case from India was reported by Sinha A *et al*. from a patient with subungual globular swelling on fifth toe.[2] As the name suggest it has a predilection for finger and toenails of middle aged adults with a male predominance. Excision is the mainstay of treatment and local recurrences are noted in upto 22% of cases.[3] Malignant behavior or metastasis has not been reported yet.

Superficial acral fibromyxoma are solitary soft tissue tumors which present in subungual and periungual region with variable size ranging from 0.6 to 5 cm in maximum dimension. It is a well circumscribed, polypoid, non-encapsulated tumor with cut section revealing off white gelatinous surface. Histopathological examination reveals the presence of hyperplasia and hyperkeratosis in epidermis. In dermis there is widespread presence of stellate and spindle-shaped fibroblast like tumor cells with myxoid or collagenous

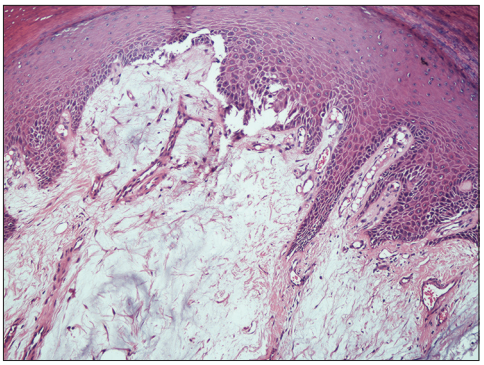

Figure 2: Histopathology of the papule showing dermal tumor composed of bland spindle and stellate cells within a myxoid or collagenous stroma with prominent vessels and mast cells (H & E, ×10)

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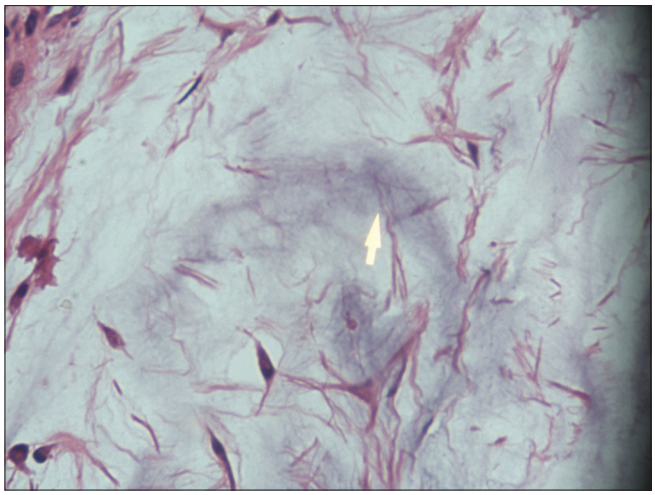

Figure 3: Bland spindle and stellate cells arranged in random loose storiform pattern (H&E, ×40)

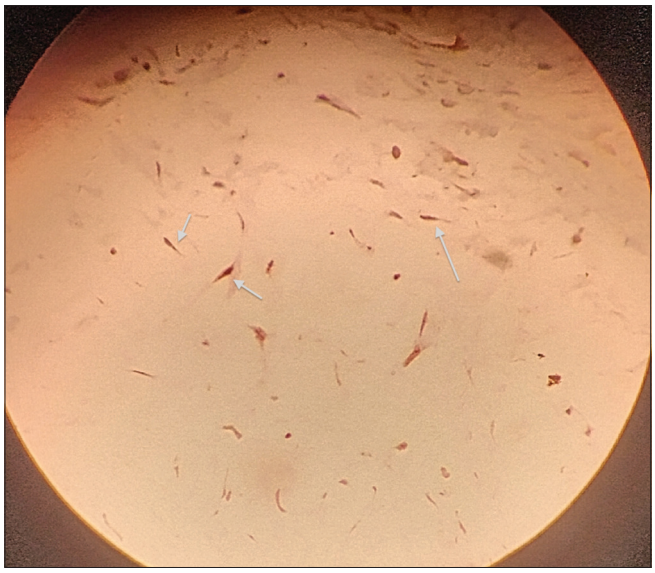

Figure 5: CD99 marker showing positivity in stellate cells on Immunohistochemistry (×100)

matrix. Blood vessel proliferation and mast cells are present throughout the dermis with mild nuclear atypia. No other type of inflammatory infiltrate seen. The margins of tumors have been described as lobulated pushing to irregular infiltrative. Alcian blue can be used to highlight the myxoid matrix and immunohistochemistry tumor cells stains for CD99, CD34, and vimentin. On electron microscopy it was found to be composed of cytoplasmic intermediate filaments and numerous cisternae of rough endoplasmic reticulum which is consistent with the fibroblastic nature of the tumor.[4]

The differential diagnosis which we kept in our case was acral digital fibrokeratoma and acral fibromyxoma. It can be differentiated from dermatofibrosarcoma protuberans, superficial angiomyxoma, glomus tumor,

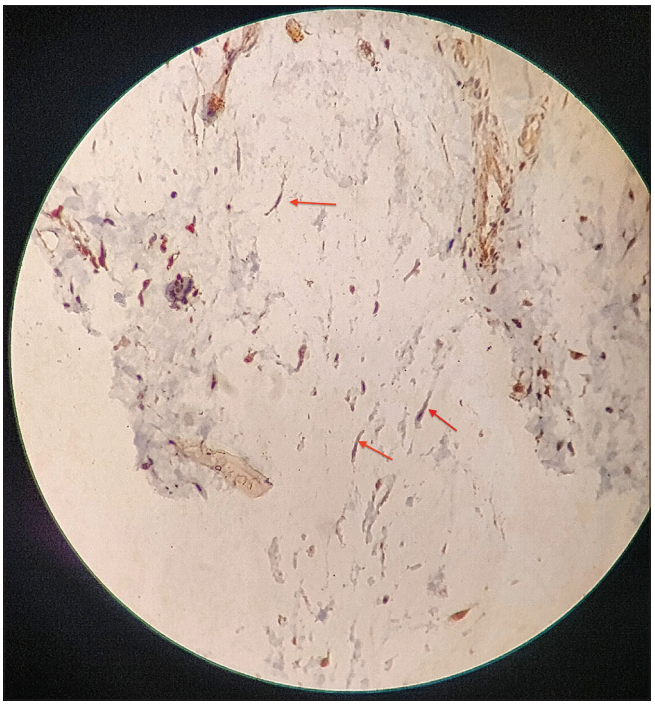

Figure 4: CD99 marker showing positivity in stellate cells on Immunohistochemistry

giant cell tumor of tendon sheath, fibrous histiocytosis and acral fibrokeratoma on the basis of histopathology and immunohistochemistry.

To conclude, superficial acral fibromyxoma is a benign soft tissue tumor of fibroblast origin with no local invasion and can be easily diagnosed on histopathological examination. Excision remains the mainstay of treatment and patient to be followed-up in future for any local recurrence.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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