Quiz

A solitary hyperkeratotic papule on the palm

A 42-year-old woman presented to us with an asymptomatic lesion on her left palm. It was present since three years and was gradually increasing in size. Clinical examination revealed a well-defined papule measuring 1 cm × 0.8 cm, surrounded by an epidermal collarette [Figure 1]. Dermoscopy showed a hyperkeratotic yellowish papule without any vascular pattern [Figure 2]. The patient was earlier diagnosed as verruca vulgaris and was treated with cryotherapy on three occasions. Due to absence of clinical improvement, an excisional biopsy was done. Histopathological examination showed spindle shaped and stellate cells in a fibromyxoid background stroma, with no obvious architectural growth pattern [Figure 3]. There were no signs of mitotic activity or nuclear pleomorphism. Immunohistochemical analysis revealed CD34 positive cells, while other antigens such as desmin, S100 and epithelial membrane antigen were negative [Figure 4].

WHAT IS YOUR DIAGNOSIS?

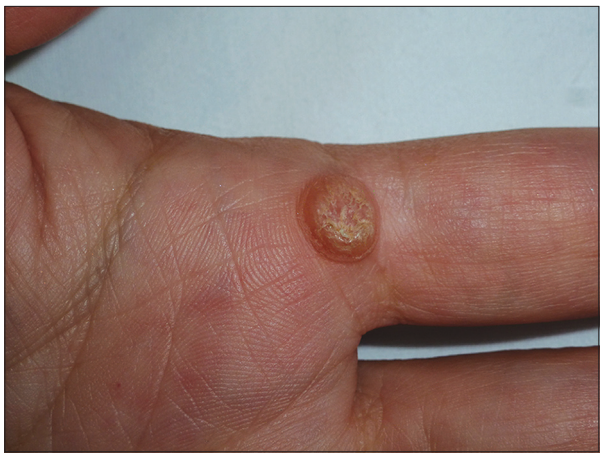

Figure 1: Well-defined papule measuring 1 cm × 0.8 cm surrounded by an epidermal collarette

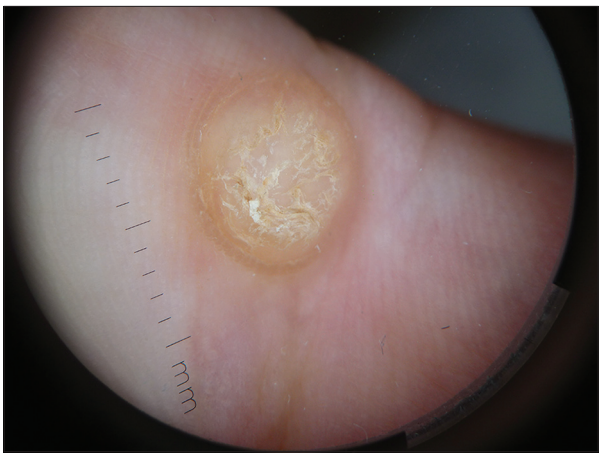

Figure 2: Hyperkeratotic yellowish papule without any vascular pattern on dermoscopy

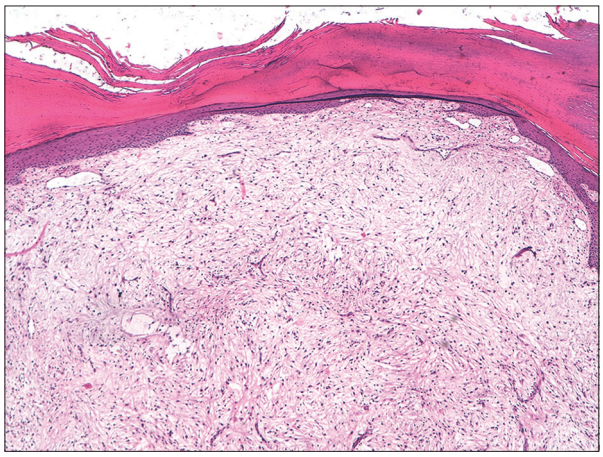

Figure 3: Spindle and stellate cells in a fibromyxoid stroma that did not follow any architectural growth pattern (H and E stain, ×400)

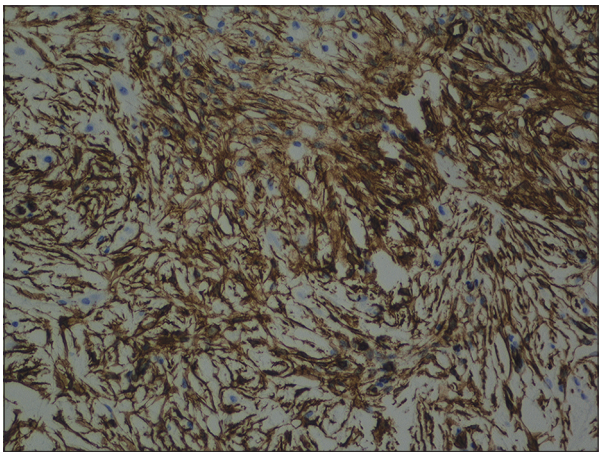

Figure 4: Immunohistochemistry showing positivity for CD34 (×400)

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ANSWER

Superficial acral fibromyxoma.

DISCUSSION

Superficial acral fibromyxoma is a rare tumor which was first described by Fetsch *et al*. in 2001.[1] They analyzed 37 patients and described the clinicopathological and immunohistochemical features. Although recently described, this entity is generally underdiagnosed.

Superficial acral fibromyxoma is a tumor that usually affects middle-aged adults with a male predilection. The most common locations are the subungual or periungual regions of the fingers and toes while the palms are usually spared. They are frequently reported to be painful but may be asymptomatic. The typical presentation is a slow growing, well defined solitary papule that is hard to rubbery in consistency. The nail apparatus may be affected in approximately 50% of patients.[2]

Dermoscopy has been mentioned as a tool to diagnose superficial acral fibromyxoma.[3] A central scar-like patch and arborizing vessels are described. Our case presented none of these findings, instead a hyperkeratotic yellowish papule with no vascular pattern was seen. As the authors described the dermoscopic appearance of only one lesion, we believe that these findings may not be applicable to all superficial acral fibromyxomas and more studies are needed.

The histopathological findings in our case were similar to the ones already described in the literature. A non-encapsulated tumor composed of spindle and stellate cells are observed within a background of fibromyxoid stroma. Nuclear pleomorphism or mitotic figures are rare. Multinucleated cells, necrotic areas and epidermal signs of viral infection have been described. Thus, the involvement of human papillomavirus has been proposed.[2,4] Immunohistochemical analysis of tumor cells show positivity for CD34. This is an essential requirement to make a differential diagnosis with myxoid dermatofibrosarcoma protuberans. myxoid dermatofibrosarcoma protuberans however presents translocation t17;22, a storiform pattern and infiltration of the subcutaneous tissue. CD34 positivity suggests a possibility of pluripotent mesenchymal origin. Actin, desmin, cytokeratin, apolipoprotein D and HMB45 are usually negative. Other myxoid spindle cell neoplasms that should be considered in the differential diagnosis include benign (myxoid neurofibroma, superficial angiomyxoma, mucous cyst) and malignant neoplasms (acral myxoinflammatory fibroblastic sarcoma, fibromyxoid sarcoma, myxofibrosarcoma). Acral neoplasms such as sclerosing perineurioma, periungual fibroma, digital fibrokeratoma and cellular digital fibroma are other differentials.

No evidence of malignant transformation or metastatic disease has been reported in our case till date. The maximum depth of invasion is usually only up to the dermis and in some cases, the subcutaneous tissue. However, there are a few cases in the literature that describe bone extension.[4] No distant metastases have been described.[5]

The management of superficial acral fibromyxoma is surgical excision with tumor free margins. There may be local recurrences if surgical excision is incomplete.[5] We recommend regular follow-up of these patients to detect recurrences early.

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Conflicts of interest
There are no conflicts of interest.

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