Dermpath Quiz

Johanna Durda[1,2], Viktoryia Kazlouskaya[2], Elen Blochin[2], Ciara A. Maguire[2], Dirk Elston[2]

[1]Department of Dermatology, Elbe Klinikum Buxtehude, Buxtehude, Germany, [2]Ackerman Academy of Dermatopathology, New York, USA

A 35-year-old female patient with a several month history of a skin-colored nodule on the toe presented to the dermatology clinic. Clinically, the lesion was suggestive of a fibroma.

Histopathological sections showed a dermal tumor composed of stellate and spindle-shaped cells with fasciculated growth between collagen bundles and a myxoid stroma [Figures 1 and 2]. The deep portion of the lesion demonstrated a nodular growth pattern with some infiltration of surrounding collagen, myxoid stroma and mast cells. Immunohistochemical stains demonstrated diffuse strong expression of CD34 [Figure 3] and focal positivity for CD 99. Only scattered Factor XIIIa cells were present. S100 and EMA were negative.

The tumor most likely represents:

A) Acquired digital fibrokeratoma
B) Periungual fibroma
C) Digital fibromyxoma
D) Neurofibroma
E) Inclusion body fibroma

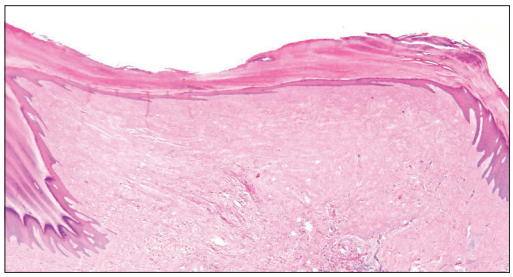

Figure 1: Polypoid lesion of acral skin. [H & E stain, ×20]

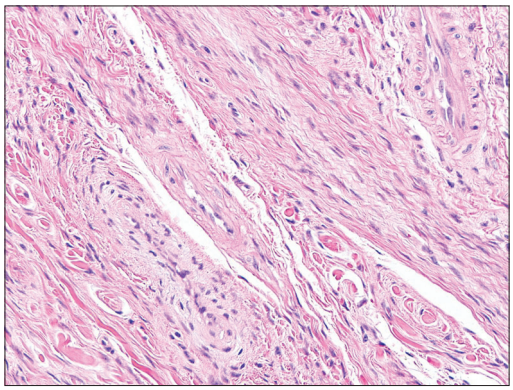

Figure 2: Fasciculated growth of spindle cells between collagen bundles in the superficial portion of the lesion. [H & E, ×400]

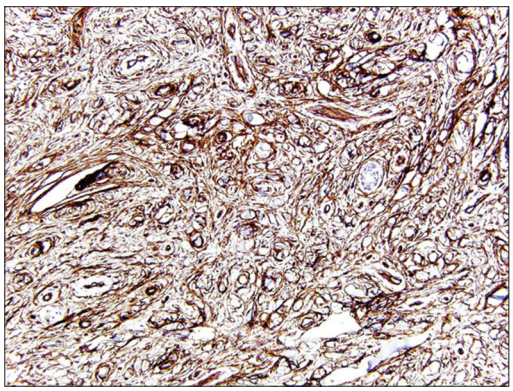

Figure 3: Diffuse expression of CD34, magnification ×20

Access this article online
Website: www.idoj.in
DOI: 10.4103/2229-5178.115536
Quick Response Code:

Address for correspondence:
Dr. Dirk Elston,
145 East 32nd St
10th Floor, New York,
USA.
E-mail:
delston@ameripath.com

ANSWER: C. Digital fibromyxoma

DISCUSSION

Presence of a nodular dermal component with mucinous stroma, spindle- or stellate-shaped cells within a loose myxoid stroma, focal infiltration of the dermis and positivity with CD34 all serve as clues to the diagnosis of digital fibromyxoma (superficial acral fibromyxoma).

Digital fibromyxoma is a benign soft tissue tumor with a nodular myxoid or cellular component and a predilection for acral sites. It typically has a loose storiform and fascicular growth pattern and the cells are surrounded by a myxoid or collagenous stroma. The tumor itself has a striking predilection for the fingers and toes, but occurrence on other portions of the hands and feet including the heel has been reported.[1] Nuclear atypia and mitotic figures are rarely seen. But multinucleate stromal cells are occasionally noted.[2]

The superficial portion of the tumor can resemble acquired digital fibrokeratoma, but in contrast to acquired digital fibrokeratomas and periungual fibromas, there is a deep nodular component, typically with infiltration of surrounding collagen. In addition, digital fibromyxoma has a less tendency for concentric perivascular fibrosis and vertical streaking of collagen, especially in the deeper component [Figure 3]. In digital fibromyxomas, an overlying keratin horn and collarette are typically lacking, Factor XIIIa is negative or only focally positive, and EMA can sometimes be expressed focally. Unlike periungual fibromas, there is no association with tuberous sclerosis. Unlike neurofibroma, S100 is negative or only weakly positive. Digital fibromyxomas lack the intersecting fascicles and SMA+ cytoplasmic inclusion bodies found in inclusion body fibroma (recurrent infantile digital fibroma).

Digital fibromyxoma was first described by Fetsch *et al*. in 2001 as superficial acral fibromyxoma.[3] Prescott *et al*, reviewed a series of 771 tumors from acral sites diagnosed between 1970 and 2006. Forty-one cases were reclassified as digital fibromyxoma, suggesting that the tumor is frequently misclassified.[4]

This neoplasm can present at a wide range of ages (from 4 to 91), but most patients are middle-aged with some male predominance. The size of the lesion ranges from 0.5 to 5.0 cm. Dermal infiltration is common, but bone invasion is rare.[5,6] Simple excision is usually adequate for recurrent tumors and malignant degeneration has not been reported, but recurrence after excision has been reported in up to 24% of cases.

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Cite this article as: Durda J, Kazlouskaya V, Blochin E, Maguire CA, Elston D. Dermpath Quiz. Indian Dermatol Online J 2013;4:244-5.

Source of Support: Nil, **Conflict of Interest:** Nil.