Slow-growing thumb nodule in an African American female

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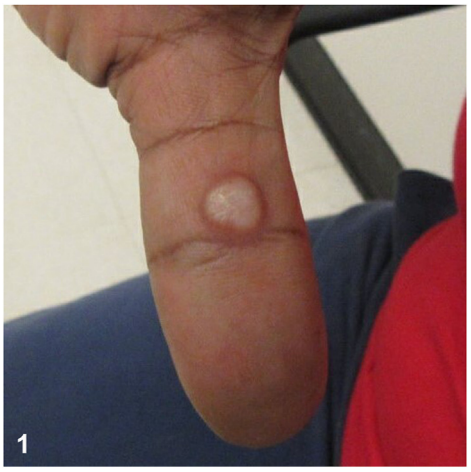

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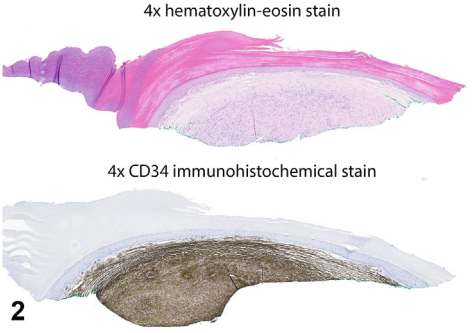

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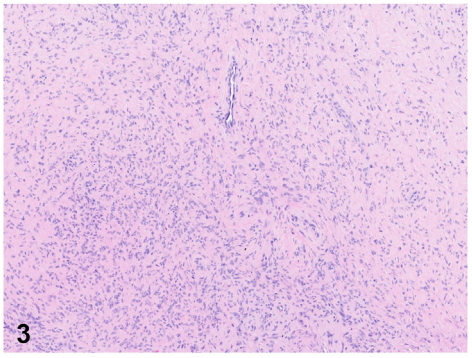

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CASE VIGNETTE

A 61-year-old African American female presented with a 1-year history of an asymptomatic bump on her right thumb that had been gradually increasing in size. Examination revealed a firm, well-circumscribed, smooth, pale white nodule with peripheral erythema on the ventral aspect of the right thumb just proximal to the interphalangeal joint (Fig 1). Radiograph of the hand did not demonstrate osseous extension. Histopathologic examination showed diffuse reticular dermal involvement by densely packed haphazardly arranged $CD34^+$ short spindle cells, with few interspersed banal-appearing vessels and slight thinning of overlying epidermis. (Fig 2: hematoxylin-eosin, 4×; CD34 immunohistochemical stain, 4×; Fig 3: hematoxylin-eosin, 10×).

Question 1: What is the most likely diagnosis?

A. Superficial acral fibromyxoma

B. Cellular digital fibroma

C. Sclerotic fibroma

D. Digital mucous (myxoid) pseudocyst

E. Digital fibrokeratoma

Answer:

A. Superficial acral fibromyxoma — Incorrect. Superficial acral fibromyxomas are slow-growing myxoid tumors and histologically characterized by stellate cells in a myxocollagenous matrix with a poorly circumscribed margin. No significant myxoid stroma is noted in this case.

B. Cellular digital fibroma — Correct. A cellular digital fibroma is a benign, fibrous nodule seen most commonly in an acral distribution. It can be distinguished clinically from a dermatofibroma by a lack of a "dimple sign" and from an acral fibrokeratoma by an absence of a surrounding collarette of skin. On histology, there is a dermal-based cellular proliferation of haphazardly arranged bland spindle cells.[1,2]

C. Sclerotic fibroma — Incorrect. A sclerotic fibroma, also known as a circumscribed storiform collagenoma, is an uncommon skin nodule characterized histologically by a relatively hypocellular lesion with prominent sclerotic collagen bundles imparting a characteristic "plywood" appearance.

D. Digital mucous (myxoid) pseudocyst — Incorrect. A digital mucous pseudocyst is a shiny papule found at the end of fingers/toes and without a true capsule (hence the term "pseudocyst"). It is characterized by degeneration of connective tissue overlying the last segment of the fingers/toes (usually <1 cm away from the nail) leading to extravasation of synovial fluid. This gives them a characteristic shiny, semitranslucent appearance.

E. Digital fibrokeratoma — Incorrect. A digital fibrokeratoma appears as a solitary, firm raised area on the skin on the acral surfaces. There is often a collarette of scale surrounding the base of the lesion and an overlying firm thickening of skin. Histologically, it is characterized by a rim of epidermis which forms a collarette and a thick core of vertically oriented collagen surrounded by fine capillaries and connective tissue present at the center of the lesion without significant increase in dermal cellularity.

Question 2: Which immunohistology marker is most strongly associated with a cellular digital fibroma?

A. Vimentin

B. Epithelial membrane antigen

C. CD34

D. Cytokeratin AE1/AE3

E. SRY-related HMG-Box gene 10

Answer:

A. Vimentin — Incorrect. Vimentin is constitutively expressed in mesenchymal cells. While it is typically positive in a cellular digital fibroma,[1] it is nonspecific and positive in a variety of other mesenchymal tumors.

B. Epithelial membrane antigen — Incorrect. Epithelial membrane antigen is a marker that's used to distinguish neoplasms of epithelial origin. It is usually positive in superficial acral fibromyxoma.

C. CD34 — Correct. CD34 is human hematopoietic progenitor cell antigen that is strongly associated with cellular digital fibroma. This type of lesion shows both diffuse and strongly positive staining with this marker and is a pathognomonic feature of this tumor. Although dermatofibrosarcoma protuberans is also strongly associated with CD34,[3] it demonstrates deeper extension into the subcutaneous fat, whereas cellular digital fibroma is well circumscribed and not as deeply infiltrating as in our case.

D. Cytokeratin AE1/AE3 — Incorrect. Cytokeratin AE1/AE3 is a mixture of 2 clones of antibodies used to detect both high- and low-molecular-weight

keratins. It is commonly used as a marker for neoplasms of epithelial origin.

E. SRY-related HMG-Box gene 10 — Incorrect. SRY-related HMG-Box gene 10 is an important nuclear transcription factor that plays a role in the development of neural crest cells as they differentiate into melanocytes. It is a sensitive and specific marker for melanocytic lesions.

Question 3: If treatment is sought, what is the best next step in management?

A. Cryoablation

B. Shave excision

C. Full-thickness excision

D. Electrodessication and curettage

E. Photodynamic therapy

Answer:

A. Cryoablation — Incorrect. Cryoablation is commonly used for the treatment of warts, actinic keratoses, seborrheic keratoses, and molluscum contagiosum. Due to the nodule size and depth, it is not the ideal choice for a cellular digital fibroma.

B. Shave excision — Incorrect. A shave excision has the advantage of not requiring sutures; however, it is typically limited for lesions that predominate in the upper layers of the skin, unlike a cellular digital fibroma.

C. Full-thickness excision — Correct. A full-thickness excision including the epidermis and dermis is the best choice for a cellular digital fibroma. This will ensure complete removal and minimize the chance for recurrence, although the risk of recurrence is extremely low.

D. Electrodessication and curettage — Incorrect. Electrodessication and curettage, similar to a shave excision, is limited for lesions that predominate in the upper layers of the skin, unlike a cellular digital fibroma. Additionally, broad superficial lesions in areas with thick underlying dermis, such as the trunk and extremities, are ideal candidates for electrodessication and curettage.

E. Photodynamic therapy — Incorrect. Photodynamic therapy involves targeted topical application of a photosensitizer combined with light energy and has no known role in the treatment of cellular digital fibroma. It is typically reserved for precancerous lesions and nonmelanoma skin cancer.

Abbreviations used:

EMA: Epithelial membrane antigen
CD34: Cluster of Differentiation 34
ED&C: Electrodessication & Curettage
PDT: Photodynamic therapy

Conflicts of interest

None disclosed.

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