Dermatofibrosarcoma Protuberans Arising in a Digit: A Case Report

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Introduction

- **Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing, highly infiltrative soft tissue tumor with low metastatic potential.**
- Usually located on the trunk or extremities, but rarely the digits
- Staining and cytogenetic studies are critical for accurate diagnosis
- **We report a case of DFSP on the digit of a 43-year-old male that presented initially as an atypical CD34 positive tumor.**

Case report

- 43-year-old male presented with a painless well-circumscribed slow-growing nodule on the dorsum of the third digit for 1 year.
- Ddx: cellular digital fibroma, rheumatoid nodule, cholesterol nodule and giant cell tumor of the tendon sheath.
- Pathologic findings:
  - Whorled spindle cell proliferation in a storiform pattern within the dermis [Fig 1 & 2]
  - Diffuse CD34 positive staining in tumor cells [Fig 3]
  - Patchy positive staining for EMA
  - Negative for Factor XIIIa, SOX-10 and STAT6

Case Report (Continued)

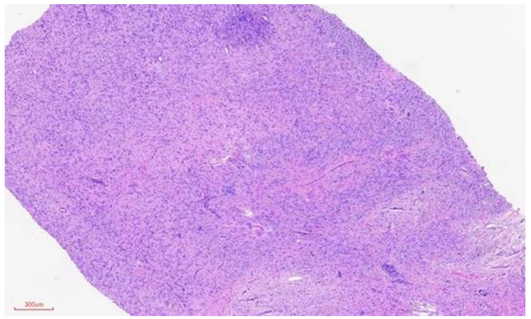

Figure 1: H&E stain at 40x showing spindle cell proliferation in the dermis.

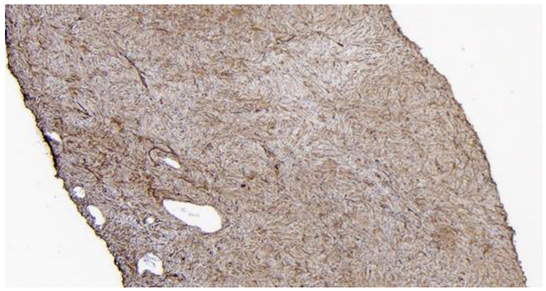

Figure 2: Immunohistochemistry stain at 40x showing diffuse positivity for CD34

- **FISH analysis showed an abnormal pattern of the PDGFB (22q13.1) locus**
- **Diagnosed with DFSP based on clinical, histologic, and cytogenetic findings.**
- Referred to Mohs micrographic surgery for prompt removal.

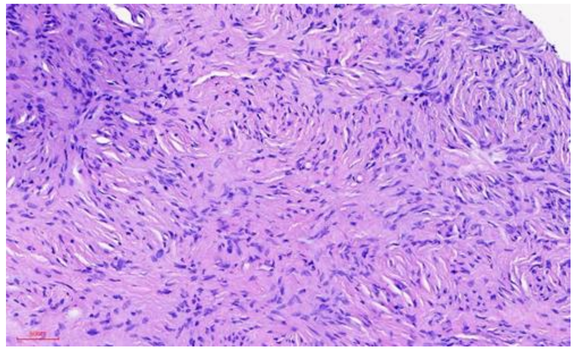

Figure 3: H&E stain at 200x showing characteristic spindle cells arranged in a whorling storiform pattern with elongated nuclei.

Conclusions

- **This case is one of only a handful of reported cases of DFSP presenting on the digit.**
- The COL1A1-PDGFB translocation contains the chromosomal abnormality t(17;22)(q22;q13), which contributes to the pathogenesis of this rare soft tissue sarcoma [1].
- With equivocal staining, cytogenetic analysis for this fusion gene can be used to exclude a diagnosis of DFSP in patients with overlapping features, such as with a CD34+ dermatofibroma [2].
- Because of the aggressive nature of this tumor, Mohs surgery with histologic marginal overlook is performed. Occasionally, partial amputation may be necessary due to limited availability of tissue for preservation [3].
- **It is crucial that cytogenetic studies be performed on digital slow-growing nodules to differentiate between a clinically similar benign lesion, specifically CD34+ cellular digital fibroma, and a malignant DFSP.**

References & Disclaimers

1. Hao X, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, Vidimos AT. Dermatofibrosarcoma Protuberans: Update on the Diagnosis and Treatment. J Clin Med. 2020 Jun 5;9(6):1752. doi: 10.3390/jcm9061752. PMID: 32516921; PMCID: PMC7355835.
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3. Do AN, Goleno K, Geisse JK. Mohs micrographic surgery and partial amputation preserving function and aesthetics in digits: case reports of invasive melanoma and digital dermatofibrosarcoma protuberans. Dermatol Surg. 2006 Dec;32(12):1516-21. doi: 10.1111/j.1524-4725.2006.32366.x. PMID: 17199665.

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