Aggressive Digital Papillary Adenocarcinoma

Jessica Thomas MD, Alexandra Thomson MD, Savannah Grunhard MD, George Negrea MD

Background

- Aggressive digital papillary adenocarcinomas are derived from sweat glands, originating from the apocrine and eccrine glands
- Since these are often painless masses and not associated with constitutional symptoms, it can take months or years before a patient will seek medical attention
- These tumors are often under-recognized and not biopsied, which can be fatal to patients as there is a high potential for metastasis
- This rare type of malignancy was first discovered in 1979 and to our best knowledge, there are only approximately 200 cases reported since

Clinical Presentation

This patient is a 72-year-old male who presented to the oncology office as a referral for a rare form of skin malignancy. Patient stated that he noticed a nodule on the volar aspect of the right thumb about 3 years ago. It grew slowly over time, and he went to a dermatologist for evaluation. He subsequently underwent extensive debridement but a few months later, the lesion grew back. It was then re-evaluated and believed to be possibly a ganglion cyst, so it was fully excised. Pathological examination of the specimen revealed a 1.5 x 0.7 x 0.4 cm fragment of tan-pink fibroconnective tissue. A 0.7 cm disrupted thin-walled cystic space was identified. It was histologically read as digital papillary adenocarcinoma with increased mitotic activity and accompanying single cell necrosis. The patient then underwent a deep resection of the tumor bed. Decision was made to schedule a PET-CT scan to rule out metastasis and the patient was presented at the local tumor board for additional guidance. Fortunately, PET-CT did not show any evidence of metastatic malignancy. However, after thorough review of limited case studies, it was recommended that the patient would require an additional wider excision or a complete distal right thumb amputation to avoid local recurrence. The radiation oncologists felt there was not enough information currently on how effective radiation is for this rare type of malignancy. Ultimately, the patient was able to have thumb sparing surgery and continues to have annual PET-CT scans.

Pathology

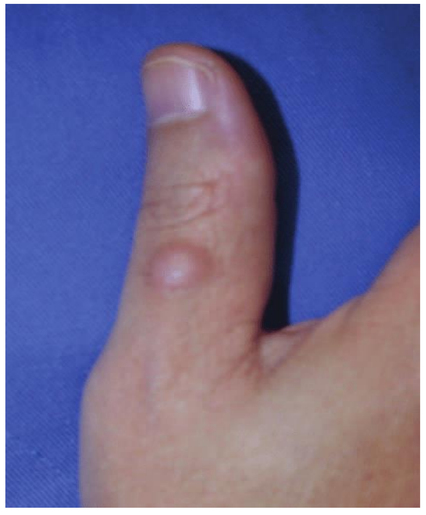

Figure 1. Digital papillary adenocarcinoma of the thumb https://link.springer.com/article/10.2165/00128071-200708040-00007

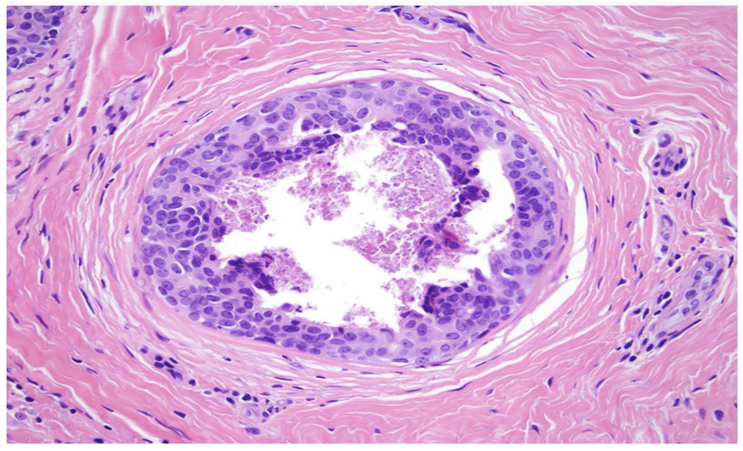

Figure 2. Histopathology of digital papillary adenocarcinoma, demonstrating cellular atypia, nuclear contour irregularity, and focal necrosis
https://dermpath.weill.cornell.edu/diagnosis-gallery/digital-papillary-adenocarcinoma

Discussion

- The differentials are, but not limited to, include soft tissue infections, ganglion cyst, foreign body granulomas, hemangiomas, calluses, or gout
- Wide local excision or digital amputation is the current treatment of choice
- There are no formal oncologic guidelines on how to diagnose, work up, treat, or follow-up patients with this type of cancer
- The current recommendations are based on case reports
- The use of chemotherapy or radiation remains unclear
- This patient only required wide local excision and thumb-sparing surgery; he continues to have normal PET-CT imaging. However, the treatment would be more aggressive if there was evidence for metastasis

Conclusion

- This case report supports full excision of suspicious lesions with additional wide margins in the setting of local disease
- Hopefully in the future, radiation or immunotherapy could target these rare tumors without the need for surgery and possible amputation

References

1. Carter K, Yao JJ, Melton SD, Lopez J, Huerta S. Digital Papillary Adenocarcinoma: Presentation, Natural History and Management. Rare Tumors. 2015 Jun 25;7(2):5867. doi: 10.4081/rt.2015.5867. PMID: 26266016; PMCID: PMC4508647.
2. Vale C, Carvalho A, Roseiro T, Antunes J, Tellechea Ó, Oliveira V: Digital Papillary Adenocarcinoma, a Rare Malignant Tumor. Case Rep Orthop Res 2019;2:89-93. doi: 10.1159/000504126
3. Nishinda H. Digital papillary adenocarcinoma. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/skintumornonmelanocyticaggressivedigital.html. Accessed October 9th, 2022.
4. Rismiller K, Knackstedt TJ. Aggressive Digital Papillary Adenocarcinoma: Population-Based Analysis of Incidence, Demographics, Treatment, and Outcomes. Dermatol Surg. 2018 Jul;44(7):911-917. doi: 10.1097/DSS.0000000000001483. PMID: 29578881.
5. Kobayashi, Tadahiro MD, PhD*,†; Hiura, Azusa MD*; Oishi, Kyosuke MD, PhD*,†; Maeda, Shintaro MD, PhD*,†; Le Pavoux, Andres James MD*; Ohara, Kuniaki MD*; Uruga, Hironori MD‡. Aggressive Digital Papillary Adenocarcinoma With Multiple Organ Metastases: A Case Report and Review of the Literature. The American Journal of Dermatopathology: December 2016 - Volume 38 - Issue 12 - p 910-914 doi: 10.1097/DAD.0000000000000703
6. Surowy HM, Giesen AK, Otte J, Büttner R, Falkenstein D, Friedl H, Meier F, Petzsch P, Wachtmeister T, Westphal D, Wieczorek D, Wruck W, Adjaye J, Rütten A, Redler S. Gene expression profiling in aggressive digital papillary adenocarcinoma sheds light on the architecture of a rare sweat gland carcinoma. Br J Dermatol. 2019 May;180(5):1150-1160. doi: 10.1111/bjd.17446. Epub 2019 Jan 20. PMID: 30472730.
7. Magro, C. M., Ma, L., & Wanf, X. (n.d.). *Digital papillary adenocarcinoma*. Dermatopathology. Retrieved March 30, 2023, from https://dermpath.weill.cornell.edu/diagnosis-gallery/digital-papillary-adenocarcinoma

- Chi, C.-C., Kuo, T.-tong, & Wang, S.-H. (2007). Aggressive digital papillary adenocarcinoma. *American Journal of Clinical Dermatology*, *8*(4), 243–245. https://doi.org/10.2165/00128071-200708040-00007

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.