A case of sweat-gland carcinoma with neuroendocrine differentiation

Shigeru Koizumi,¹ Yaei Togawa,² Inozume Takashi,² Michiyo Nakano¹

¹Division of Dermatology, Asahi General Hospital, Asahi, ²Department of Dermatology, Chiba University Hospital, Chiba, Japan

Dear Editor,

Low-grade neuroendocrine tumors, also known as carcinoid tumors, are slow-growing cancers derived from neuroendocrine cells, usually found in the digestive tract or lung lining; the primary tumor in the skin is extremely rare and is called sweat-gland carcinoma with neuroendocrine differentiation (SCAND) as a new disease concept.¹⁴ Herein, we report a case of SCAND on the lower abdomen presenting bilateral inguinal lymph node metastasis.

A 73-year-old Japanese male patient presented to our hospital with a 5-year history of a red nodule arising from the lower abdomen to the base of the penis. He has a medical history of hypertension, atrial fibrillation, and cerebral infarction. Physical examination revealed a red mulberryshaped nodule with red papules (Figure 1A) and enlarged, elastic, and complex lymph nodes at the bilateral inguinal region. Dermoscopy showed branched and/or partially reticular vessels on a pale red structureless area (Figure 1B). Contrastenhanced computed tomography revealed two tumors in the left kidney. They were renal cell carcinomas classified as cT1aN0M0 stage I with a low probability of metastasis, and no other visceral malignancies were found. 18F-fluorodeoxyglucose positron emission tomography integrated with computed tomography showed abnormally increased cellular uptake of glucose on the lower abdomen and bilateral inguinal lymph nodes; however, the left renal tumors and other sites showed normal uptake. Skin biopsy revealed various-sized tumor nests diffusely infiltrated from dermis to subcutaneous tissue (Figure 2A-B). Focal pagetoid extension of the tumor cells within the overlying epidermis was observed. The tumor cells were small to medium size, with abundant and pale eosinophilic cytoplasm, round-to-oval nuclei, and coarse granular chromatin pattern. Few nuclear atypia and mitotic figures were observed (Figure 2C). A tumor mass floating in abundant mucus was observed in part of the tumor (<10%) (Figure 2D). Alcian blue staining (pH 2.5) showed extracellular mucin deposition. The metastatic lymph nodes were almost replaced by tumor cells that showed a trabecular growth pattern (Figure 2E). Immunohistochemical staining of the tumor cells was positive for cytokeratin 7, estrogen receptor, progesterone receptor, chromogranin A, synaptophysin, CD56, and GATA3 and negative for cytokeratin 20. Consequently, we diagnosed this tumor as SCAND. However, the scheduled surgery was canceled due to multiple cerebral infarctions. SCAND was a newly proposed entity and sporadic disease in 2022. It commonly occurs on milk-lines on the trunk of middle-aged to elderly males and often causes lymph node metastasis.⁴ In our case, histopathologically, a tumor mass floating in a large amount of mucus was observed in part, and primary cutaneous mucinous carcinoma (PCMC) with neuroendocrine differentiation was the differential diagnosis. However, since this feature accounts for only a tiny fraction of tumors (<10%) and



Figure 1. A) a red mulberry-shaped nodule on the lower abdomen to the base of the penis (30 mm \times 25 mm \times 15 mm) with some red papules on the margin; B) a dermoscopic image of the penile nodule showing branched and/or partially reticular linear vessels in a pale red structureless area.



Correspondence: Yaei Togawa, Department of Dermatology, Chiba University Hospital, 1-8-1 Inohana, Chuo-ku, Chiba-shi, Chiba, 260-8677, Japan.

Tel.: +81.432227171 - Fax: +81.432262128. E-mail: togawa-yk@faculty.chiba-u.jp

Key words: carcinoid tumor, lymphatic metastasis, mucinous carcinoma, neuroendocrine tumor.

Acknowledgment: we would like to thank Editage (www.editage.com) for English language editing.

Contributions: SK, manuscript writing; YT, IT, conception and design; YT, MN, final approval of the manuscript.

Conflict of interest: the paper is exempt from Ethical Committee approval.

Funding: none

Received for publication: 18 April 2022. Accepted for publication: 25 April 2022.

This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License (CC BY-NC 4.0).

©Copyright: the Author(s), 2023 Licensee PAGEPress, Italy Dermatology Reports 2023; 15:9513 doi:10.4081/dr.2022.9513

Publisher's note: all claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article or claim that may be made by its manufacturer is not guaranteed or endorsed by the publisher.

PCMC occurs primarily in the head and neck region, we thought PCMC could be ruled out. Consequently, while SCAND and PCMC sometimes share histologic similarities, their differentiation is important because PCMC rarely metastasizes while SCAND often does.⁵ It has not been long since the SCAND concept was proposed, and more cases are expected to accumulate.

References

- Van Dijk C, Ten Seldam RE. A possible primary cutaneous carcinoid. Cancer 1975;36:1016-20.
- Chen TY, Morrison AO, Susa J, Cockerell CJ. Primary low-grade neuroendocrine carcinoma of the skin: An



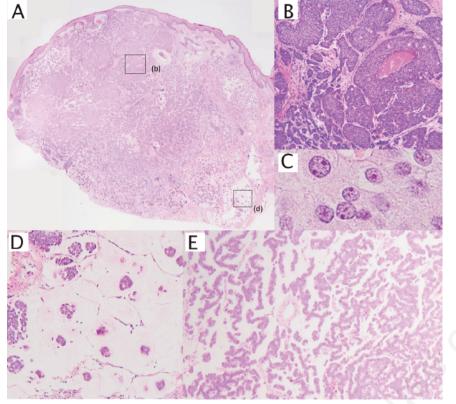


Figure 2. A) histopathological features of the primary tumor. The tumor nests show diffuse infiltration from the upper dermis to the subcutaneous tissue (hematoxylin-eosin, original magnification \times 40); B) tumor nests of varying sizes are seen (hematoxylineosin, \times 200); C) the tumor cells are small- to medium-sized with abundant and pale eosinophilic cytoplasm, round-to-oval nuclei, and a "salt-and-pepper" chromatin pattern. Few nuclear atypia and mitotic figures are observed (hematoxylin-eosin, \times 400); D) a tumor mass floating in abundant mucus is observed in part of the tumor (<10%) (hematoxylin-eosin, \times 200); E) tumor cells displayed a trabecular growth pattern with abundant extracellular mucin deposition in the resected lymph node (hematoxylineosin, \times 200).

exceedingly rare entity. J Cutan Pathol 2017;44:978-81.

- Goto K, Anan T, Nakatsuka T, et al. Low-Grade Neuroendocrine Carcinoma of the Skin (Primary Cutaneous Carcinoid Tumor) as a Distinctive Entity of Cutaneous Neuroendocrine Tumors: A Clinicopathologic Study of 3 Cases With Literature Review. Am J Dermatopathol 2017;39:250-8
- Goto K, Kukita Y, Honma K, et al. Sweat-gland carcinoma with neuroendocrine differentiation (SCAND): a clinicopathologic study of 13 cases with genetic analysis. Mod Pathol 2022;35: 33-43.
- Kim BJ, Choi HJ, Kim HJ et al. A Case of Primary Cutaneous Mucinous Carcinoma with Neuroendocrine Differentiation. Ann Dermatol 2010;22: 472-7.