Squamomelanocytic Tumor of the Nail Unit Metastasizing to a Sentinel Lymph Node: A Dermoscopic and Histologic Investigation

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Introduction
Squamomelanocytic tumor (SMT) is a true malignant cutaneous proliferation composed of closely intermingled cells of squamous cell carcinoma (SCC) and melanoma. Since the publication of a first case series in 1999 [1] only 9 more cases have been reported in the world literature [2–9]. The large majority of earlier cases (11 out of 12) involved chronically sun-exposed skin of the face and scalp area.

A lack of consensus about the proper terminology has led to an interchangeable usage of different expressions. In a more precise interpretation SMT has to be differentiated from a ‘collision’ of a well-demarcated SCC and melanoma, where the two cell populations are not truly intimately mixed. In contrast to SMT, a biphenotypic malignant proliferation [10] is characterized by tumor cells that arise from a common precursor and stain for markers of both lineages in the same tumor cells (e.g. S-100 and keratin). Moreover, SMT should not be confused with a reactive colonization of only one malignant proliferation with another cell type, e.g. pseudocarcinomatous hyperplasia in melanoma [11] or colonization of SCC with cytologically unremarkable melanocytes [12]. Noteworthy, a case with two concurrent neoplasms involving different nails of the same hand has been reported [13].

We report a first SMT case involving the nail unit of the same finger, which metastasized to the sentinel lymph node. Clinical and dermoscopic aspects, as well as results of an in-depth histopathologic investigation, are presented.

Case Report
A 75-year-old man presented with an asymptomatic dystrophy of his right thumbnail. He had first noticed changes in nail morphology 2 years earlier and was repeatedly treated for subungual viral warts. There was no family or personal history of melanoma or nonmelanoma skin cancer. The clinical examination (fig. 1a) revealed a complete dystrophy of the nail plate with a multicolored hyperkeratotic tumor (white-yellow, pink, dark-gray). The periungual skin showed a faint gray-purple discoloration and areas of prominent brown pigmentation. Hyperkeratotic cysts were macroscopically visible.

Dermoscopy of the periungual skin identified areas with irregular diffuse gray-brown pigmentation (fig. 1b, box 1) which has been described as a key criterion for acrolentiginous melanoma [14]. White-yellow keratin cysts that are usually found in

Fig. 1. Clinical and dermoscopic presentation of an SMT of the nail unit. a Complete dystrophy of the nail plate with a multicolored hyperkeratotic tumor (white-yellow, pink, dark-gray). Boxes 1–3 indicate areas of dermoscopic images, which are depicted in b. b Box 1 shows a dermoscopic view with diffuse gray-brown pigmentation and keratin cysts, box 2 depicts multiple brown dots, box 3 shows a melanocytic fibrillar pattern combined with dotted/short linear vessels.
keratinizing tumors like SCCs or seborrheic keratoses were visible along the proximal eponychium (fig. 1b, box 1). Other dermoscopic criteria clearly associated with melanocytic lesions were also present and included multiple brown dots (fig. 1b, box 2) and a melanocytic fibrillar pattern combined with dotted/short linear vessels (fig. 1b, box 3).

Histology of a punch biopsy from the fibrillar pattern area revealed melanoma in situ. Therefore, the nail apparatus was removed en bloc with wide margins. As demonstrated in longitudinal sections (fig. 2a), the tumor extended from the posterior nail fold (fig. 2b) to the epithelium of the nail plate (fig. 2d). A central large irregular bulbous protrusion was confined to the nail matrix (fig. 2c). High-power field examination revealed two distinct and closely intermingled cell populations. Anastomosing epithelial cords and strands with formation of squamous pearls were admixed with atypical epithelioid melanocytic cells grouped in small nests (fig. 2e). The synopsis of all of these findings gave rise to the diagnosis of SMT, i.e. an ulcerated acrolentiginous melanoma (fig. 2f).

This diagnosis was confirmed by immunohistochemical analyses (fig 2f–h). Melanoma cells stained positively for markers S-100, Melan-A and HMB-45. A nuclear pattern of reactivity was found for microphthalmia-associated transcription factor. Other appeared highly unlikely. In contrast to a simple colonization of SCCs with melanocytes other appeared highly unlikely. In contrast to a simple colonization of SCCs with melanocytes, a rare tumor with closely intermingled cells of invasive melanoma and SCC. To our knowledge, among the 13 cases in the world literature [1–9], this is the first reported SMT involving the nail unit (table 1).

The clinical and dermoscopic examination revealed features of advanced acrolentiginous melanoma (destruction of the nail unit, multiple colors, atypical vascular pattern, melanoma cells invading the adjacent skin in a fibrillar pattern) as well as subungual SCC (pronounced hyperkeratosis of nail bed, keratin cysts in the periungual skin). The dermoscopic features gave rise to the suspicion for a tumor combined of a melanoma and an SCC. However, a final diagnosis in melanocytic lesions suspicious for melanoma should be made by dermatopathology that still represents the diagnostic ‘gold standard’ [16].

Histologically, the tumor showed a true malignant proliferation of melanoma cells closely intermingled with SCC cells. The distinct immunohistochemical profile, with none of the tumor cells displaying overlapping markers of both lineages, supported this interpretation and ruled out a true biphenotypic malignant population from a common precursor cell [10]. Given that the two types of tumor cells were intimately admixed, a collision of two distinct tumors or formation of a metastasis of one tumor into the other appeared highly unlikely. In contrast to a simple colonization of SCCs with melanocytes [12], which are cytologically unremarkable and show small nuclei, melanocytes in our case revealed the typical architecture of an aggressive neoplastic proliferation with a malignant, epithelioid cytomorphology and a high mitotic activity. On the other hand, identification of atypical survival = 91%) [15], no complete lymph node dissection was performed after informed consent of the patient. During 24 months of follow-up according to current melanoma guidelines, no evidence of recurrence or metastases was found.

Discussion

We present clinical, dermoscopic and histologic features of an SMT, a rare tumor with closely intermingled cells of invasive melanoma and SCC. To our knowledge, among the 13 cases in the world literature [1–9], this is the first reported SMT involving the nail unit (table 1).

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Squamous cells with mitoses, hyperchromatic atypical nuclei, prominent nucleoli, and pronounced dyskeratosis were consistent with the diagnosis of SCC, ruling out pseudocarcinomatous hyperplasia, which has been described as a reactive process in melanomas [11].

Although metastasis has not been reported in most known cases of SMT, a sentinel node micrometastasis of a lesion with a Breslow thickness of 4.3 mm was described recently [2], and one of two sentinel nodes of our patient showed a subcapsular melanoma micrometastasis. Thus, although the excellent long-term prognosis of such small metastases of melanoma did not necessitate further therapeutic action in our case [15], lymph node metastasis of SMT is quite possible. Consequently, in the absence of prospective data with this rare tumor, we propose the adherence to current melanoma guidelines in cases of SMT.

Fig. 2. Histopathology and immunohistochemistry of SMT. a Longitudinal section through the completely removed nail unit. b Melanoma in situ of the posterior nail fold. HE. ×40. c Irregular bulbous projection of the collision tumor confined to the nail matrix containing neoplastic epithelial cords with prominent squamous pearls, admixed with grouped atypical melanocytes. HE. ×40. d Invasive malignant melanoma of the nail bed below the fragmented nail plate. HE. ×40. e Higher magnification detail of melanoma cells admixed with squamous pearls. HE. ×100. f HMB-45 immunoreactive atypical melanocytes scattered throughout the SCC. ×100. g Atypical melanocytes coexpress Melan-A. ×100. h Neoplastic squamous cells expressing cytokeratins. Clone MNF116. ×100.
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