

Dermoscopic assessment of vascular structures in solitary small pink lesions—differentiating between good and evil

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ABSTRACT The diagnosis of a single small pink papule poses a real challenge to the clinician, as the differential diagnosis of such lesions includes benign entities such as a neurofibroma or hemangioma, as well as aggressive and potentially fatal skin malignancies such as amelanotic melanoma or Merkel cell carcinoma (MCC). The absence of a benign vascular pattern and the presence of atypical vascular features under dermoscopy direct the clinician to proceed to histologic evaluation in order to rule out a malignant process in such lesions. The diagnosis of MCC is particularly problematic, given that this tumor usually lacks specific clinical diagnostic features. Low clinical suspicion for MCC may result in delayed diagnosis and poor outcomes. The dermoscopic features of MCC are also non-specific, most commonly including milky-red areas and linear irregular vessels. We report a patient who presented with two discrete pink papules on different digits that appeared three years apart. Dermoscopy helped to diagnose a harmless hemangioma in the first lesion, and a MCC in the latter. The malignant tumor was diagnosed and excised expeditiously, with no evidence of metastatic spread.

Introduction

Merkel cell carcinoma (MCC) is a highly malignant and uncommon neuroendocrine tumor of the skin. Polyomavirus infection and ultra-violet light exposure are known etiological factors in this cutaneous malignancy [1]. MCC has an aggressive behavior with early metastases and frequent recurrences [2]. It often

develops on chronically sun-damaged skin, particularly on the head and neck of elderly patients [3]. MCC commonly presents clinically as a rapidly growing shiny erythematous or violaceous papule, plaque or nodule. Due to its non-specific clinical appearance, MCC is frequently overlooked or misdiagnosed and it is not uncommon that the histopathologic diagnosis of MCC takes the dermatologist by surprise [3].

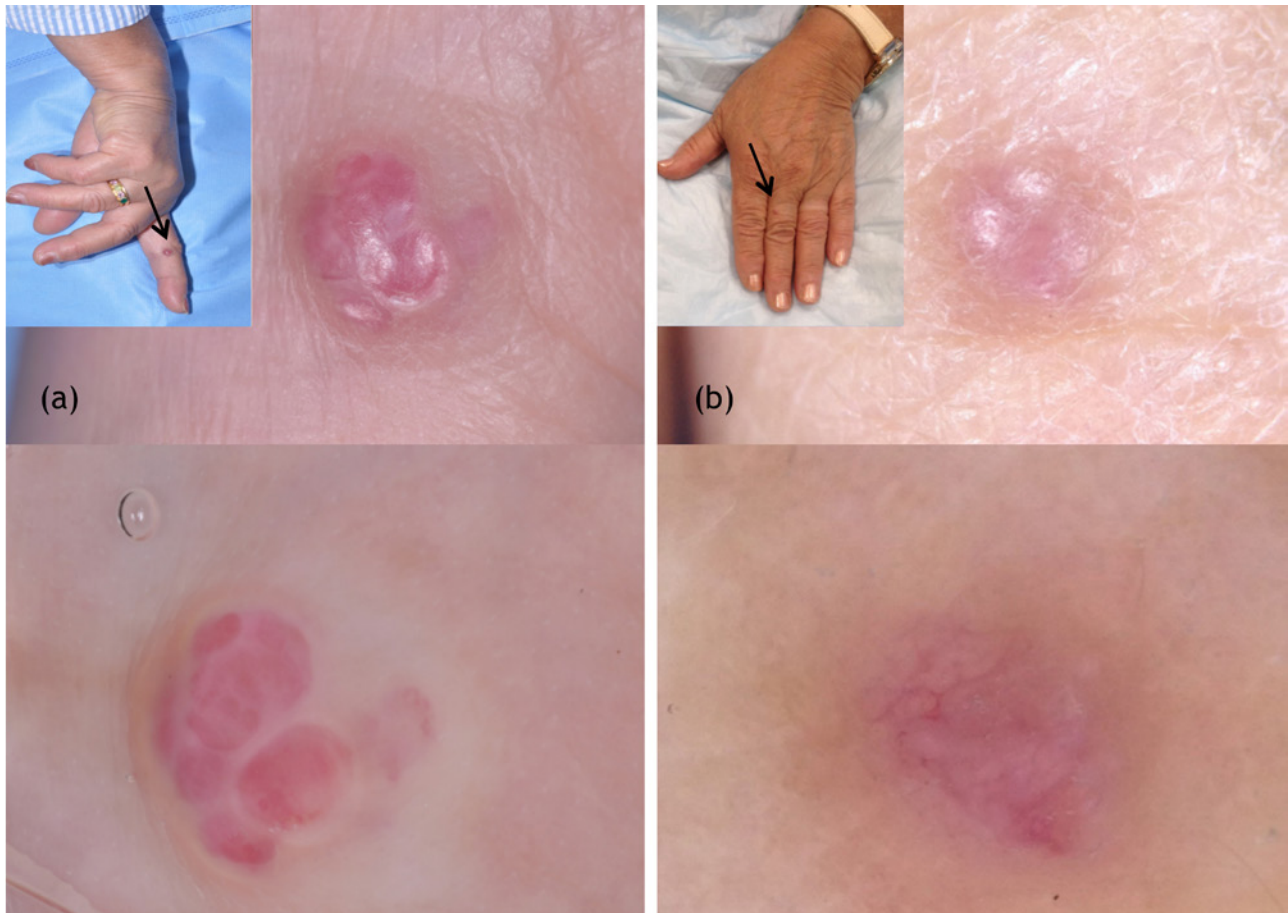


Figure 1. Clinical presentation (upper pictures) and dermoscopic appearance (lower pictures) of two digital pink papules appearing three years apart. (a) A solitary pink papule on the lateral aspect of the second digit on the right hand with red-purple lacunes under dermoscopic examination, compatible with a hemangioma. (b) A pink solitary papule on the dorsal aspect of the third finger on the left. Dermoscopy shows linear irregular blood vessels with a diffuse milky-white background. [Copyright: ©2017 Geller et al.]

Several studies have aimed to characterize the dermoscopic appearance of MCC, however no specific pattern has been identified [4-8]. Atypical vascular structures were found to be of the most diagnostic significance in MCC, most commonly linear irregular vessels and milky-red areas [6]. These dermoscopic findings are not specific and can be found in other types of cutaneous malignancies such as hypomelanotic / amelanotic melanoma [9] and melanoma metastasis [10], skin metastases from solid tumor [11], and other skin tumors [7].

Case Report

A 74-year-old Caucasian female has been followed by our clinic for more than ten years after a diagnosis in 2005 of a CD4+ small/medium pleomorphic T-cell lymphoma. In 2013 the patient presented with an erythematous papule on the second finger of her right hand. Dermoscopic examination of the lesion revealed red-purple lacuna suggestive of a hemangioma [12] (Figure 1a). Because of the patient's history of cutaneous T-cell lymphoma, a skin biopsy was performed, and the diagnosis of hemangioma was confirmed histologically. The lesion completely regressed after the biopsy was taken.

Three years later, the patient returned for another follow-up visit at which time she pointed out a new red papule on the dorsal middle digit of her left hand. Three months earlier she had noticed a small red "dot" on her finger that gradually grew in size to a 3 mm papule. On physical exam, an erythematous shiny red papule was identified. Dermoscopic examination of the papule showed linear irregular blood vessels (Figure 1b). The rest of the patient's skin was of normal appearance, and no lymphadenopathy was present. A biopsy of the lesion demonstrated a cutaneous neuroendocrine (Merkel cell) carcinoma (Figure 2). Immunohistochemical stains were positive for CK20 and Merkel cell polyomavirus, supporting the diagnosis. The patient was referred for a complete excision of the digital tumor and a sentinel lymph node biopsy. At resection there was no evidence of residual or metastatic disease, demonstrating early (stage Ia) disease [13].

Discussion

The diagnosis of a single pink papule can be extremely challenging for the clinician. Most pink papules are banal benign skin lesions. Rapid growth, ulceration, or pain are

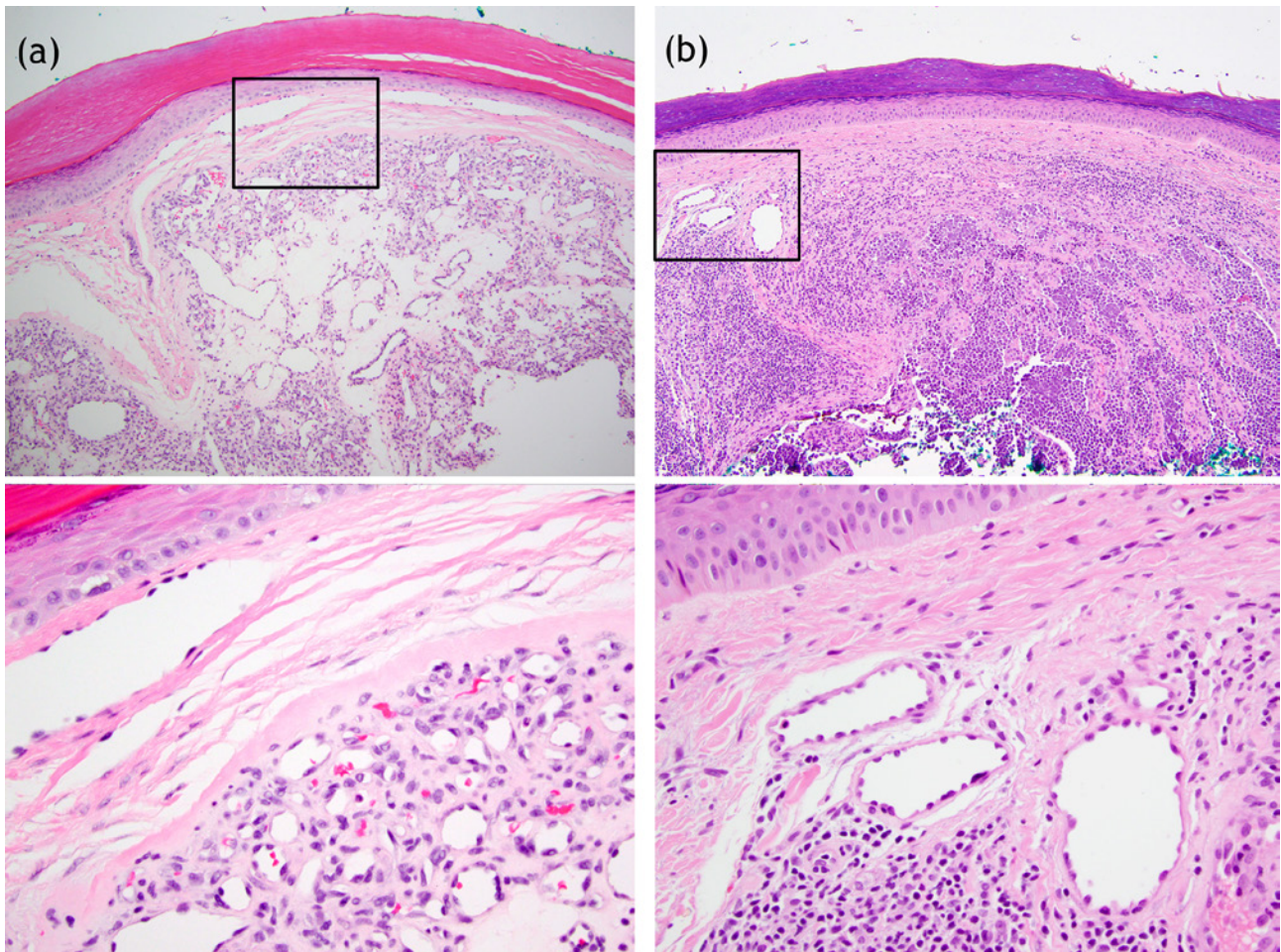


Figure 2. Histopathology of the two lesions on the fingers: (a) Pink papule on the right hand (see Figure 1a) shows large numbers of dilated vessels within the dermis compatible with capillary hemangioma. (b) Pink papule on the 3rd finger left hand (see Figure 1b) reveals dermal nodular infiltrate of small blue round-oval cells. A cluster of dilated blood vessels were noted on the upper periphery of the dermal tumor correlating with the focal linear irregular vessels seen under dermoscopy (hematoxylin and eosin stain). [Copyright: ©2017 Geller et al.]

clinical features that suggest a malignant process, though such clues are not always present. Dermoscopy is a useful diagnostic aid in approaching such lesions, as it elucidates the morphology and distribution of blood vessels and other features such as white shiny structures that may focus the differential diagnosis [14]. In small pink lesions such as a hemangioma, the diagnosis can be made by identifying the dominant vascular components of red lacunae, pale septae and milky-red areas that form a specific and diagnostic dermoscopic pattern. In other pink lesions, the presence of non-dominant, even subtle vascular structures seen on dermoscopy, when integrated with the overall morphologic appearance, may suggest a possible diagnosis. For example, the presence of glomerular vessels in an erythematous scaly lesion is suggestive of Bowen's disease [12]. In some cases, neither the gross morphology nor the dermoscopic features are diagnostically specific. In these cases, the presence of atypical blood vessels may prompt the clinician to perform a biopsy in order to rule out malignancy. Amelanotic melanoma may present as an unremarkable pink papule that may show predominantly linear irregular and dotted vascular

structures on dermoscopy as the only clue of the alarming diagnosis [9].

A linear irregular vascular pattern consists of straight vessels of irregular shape, size, and distribution. It is a well-known dermoscopic feature in skin tumors and has been described in multiple cutaneous malignancies that appear as pink or skin-colored lesions [7,9,11]. Linear irregular vessels correlate with small diameter, abnormally structured and heterogeneously dense blood vessels that are seen in the context of tumor angiogenesis [15]. In amelanotic and hypomelanotic melanoma, a predominance of linear irregular vessels on dermoscopy has been reported to have a sensitivity of 34% and a specificity of 80% for melanoma [9]. Vascular structures also predominate the appearance of amelanotic cutaneous melanoma metastases under dermoscopy, in which irregular linear vessels are identified in 45% of lesions [10]. Similarly, cutaneous metastases of solid tumors often present with a vascular dermoscopic pattern, with linear irregular vessels seen in 77% [11]. Linear irregular vessels can also be identified in other uncommon skin tumors such as adnexal tumors as well as in MCC [7,8].

The dermoscopic features of MCC have been assessed in several small case series, each involving ten to 12 MCC lesions [4-6]. The most consistent dermoscopic features in these studies were milky-red areas and linear irregular blood vessels [6]. These dermoscopic vascular features are not specific to MCC and they can be found in amelanotic melanoma and other malignant skin tumors as described above. Increased vascular density has been shown to be significantly associated with a worse prognosis in MCC as in melanoma [16]. While the clinical and dermoscopic identification of scale over a pink lesion with milky-red areas and linear irregular blood vessels could indicate a component of squamous differentiation in MCC [8], other attempts to correlate MCC dermoscopic features with other histopathologic components have failed [5]. However, in our case we show the correlation of linear irregular vessels noted on dermoscopy with the histopathological finding of clustered dilated vessels at the periphery of the tumor (Figure 2). Our case demonstrates that dermoscopic examination of MCC may provide subtle but crucial clinical clues enabling the early diagnosis of this aggressive tumor. In MCC, early detection enables expeditious treatment, and may prevent tumor progression and lead to improved survival [17].

In conclusion, dermoscopy is a helpful diagnostic tool that helps distinguish between benign and malignant tumors in patients with non-specific solitary pink lesions. The presence of linear irregular vessels in a solitary pink lesion should prompt a biopsy in order to prevent delayed diagnosis in aggressive amelanotic skin malignancies.

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