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# Clinical, dermoscopic and histological features of a Merkel cell carcinoma of the hand

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## Abstract

**Background:** Merkel cell carcinoma (MCC) is a rare and typically aggressive form of skin cancer. The benign appearance of the tumor usually on exposed skin parts, contrasting with its extensive microscopic invasion, can delay timely diagnosis.

**Main observations:** We report a case of a 71-year-old man with a slowly progressive nodule on the left hand.

**Conclusion:** At the dermoscopic examination, the presence of a polymorphous vascular pattern, including milky-red areas may constitute an additional clinical clue to accurately diagnose this rare tumor. Clinical, dermoscopic and histological features are discussed. (*J Dermatol Case Rep*. 2013; 7(1): 15-17)

## Introduction

The first description of Merkel cell carcinoma (MCC) was published in 1972 when Toker described five patients with unusual skin tumours characterized by histologically anastomosing trabeculae and cell nests.[1]

The term Merkel cell carcinoma was coined by DeWoolf-Peters in 1980 and today remains the most accepted terminology.[2]

## Case Report

A 71-year-old man was referred to our Department for a pink nodule on the back of the left hand, which had been present for 8 months. He noted that the lesions were slowly getting larger.

He had a medical history of psoriasis, benign prostatic hypertrophy and bladder carcinoma; while his family one was unremarkable.

On clinical examination, the lesion was an asymptomatic pink nodule with a soft desquamation, measuring 5,5 cm in its maximum diameter. Lymph nodes were not palpable in the cervical and axillary area.

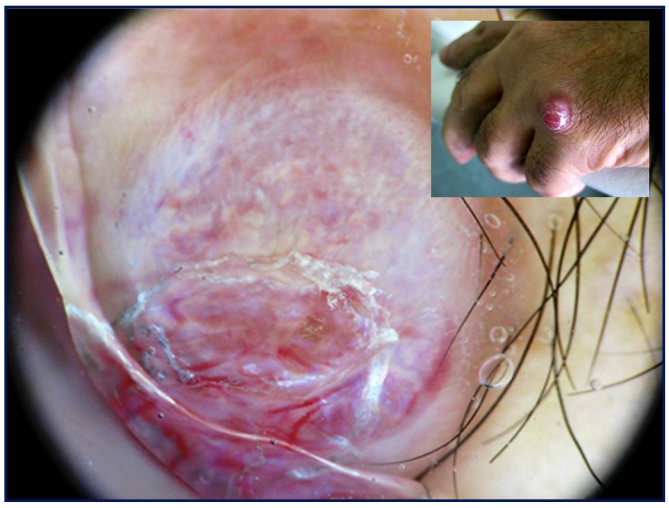

**Figure 1**

*Asymptomatic pink nodule with a soft desquamation of the left hand, measuring 5,5 cm in its maximum diameter (insert). Dermoscopically, a polymorphous vascular pattern and milky-red areas with no pigmentation can be observed.*

The dermoscopic examination revealed a polymorphous vascular pattern including milky-red areas without any pigmentation.

Based on the clinical and dermoscopic features, a tentative diagnosis of adnexal tumor was made.

The lesion was surgically excised. Histopathological examination showed a solid poorly circumscribed tumor within the dermis, composed of sheets of small round cells, characterized by hyperchromatic nuclei, without nucleoli, and scanty cytoplasm. A high rate of mitoses was present. Tumor cells were positive for cytokeratin 20 (the major epithelial marker), with a characteristic paranuclear dot-like pattern and negative for cytokeratin 7. A diagnosis of Merkel carcinoma was made.

A computed axial tomography total body was performed and resulted negative, after that the patient refused any other type of treatment or exam and was discharged from our Department.

## Discussion

MCC is a highly aggressive neuroendocrine carcinoma of the skin demonstrating a high rate of recurrence and metastasis. Indeed, 5-year rates for MCC specific survival are only about 60%.[3]

Arguments towards Merkel cell as cells of origin for MCC include that Merkel cells are the only cutaneous cells that form electron-dense neurosecretory granules present in MCC, and that Merkel cells and MCCs share similar immunophenotypic features.[3] However, several reports identified epithelial cells and sarcomatous elements in some MCCs suggesting that MCC may arise from primitive totipotent epidermal stem cells.[4] Recently, both hypotheses have been linked when Van Keymeulen and colleagues demonstrated that mammalian Merkel cells develop not from neural crest progenitors, but rather from epidermal stem cells.[5]

Probably the most important recent observation was the detection of hitherto polyomavirus, wich was named Merkel Cell Polyomavirus (MCV). Importantly, the patterns of integration suggest that MCV infection/integration occurs before the clonal expansion of the tumour cells.[3] MCV is present in about 80% of MCCs.[6]

MCCs are most often found on sun-exposed skin of Caucasian older than 50 years, with a mean age of 76 years for women and 73 years old for men at the time of initial diagnosis, with an incidence rates of 0,18-0,41 per 100 000 persons.[4]

It occurs mainly on the face and neck (40-60%), followed by the trunk (33%) and rarely on the extremities (10-20%).[7]

The primary lesion of MCC is distinguished by its absence of distinctive clinical characteristics, it presents as a rapidly growing, asymptomatic, reddish-blue dermal papule or nodule that develops over the course of weeks to months. The mnemonic AEIOU has been used to describe its clinical appearance and demographic characteristics: asymptomatic, expanding rapidly, immune suppression, older than 50 years, and ultraviolet-exposed/fair skin.[8]

The differential diagnosis must involve distinction from basocellular and squamous cell carcinoma which are common skin malignancies in parallel or skin metastasis from another primary site. Other possibilities are malignant lymphoma, keratoakanthoma and amelanotic melanoma.[9]

Dermoscopy may be helpful: in addition to dermoscopic vascular structure, often linear irregular vessels, milky-red areas are observed in more than two thirds of cases;[10] in this way, along with amelanotic melanoma and to a lesser extent BCC, the diagnosis of MCC should be included in the differential diagnosis list of a pink cutaneous nodule showing an atypical vascular pattern.

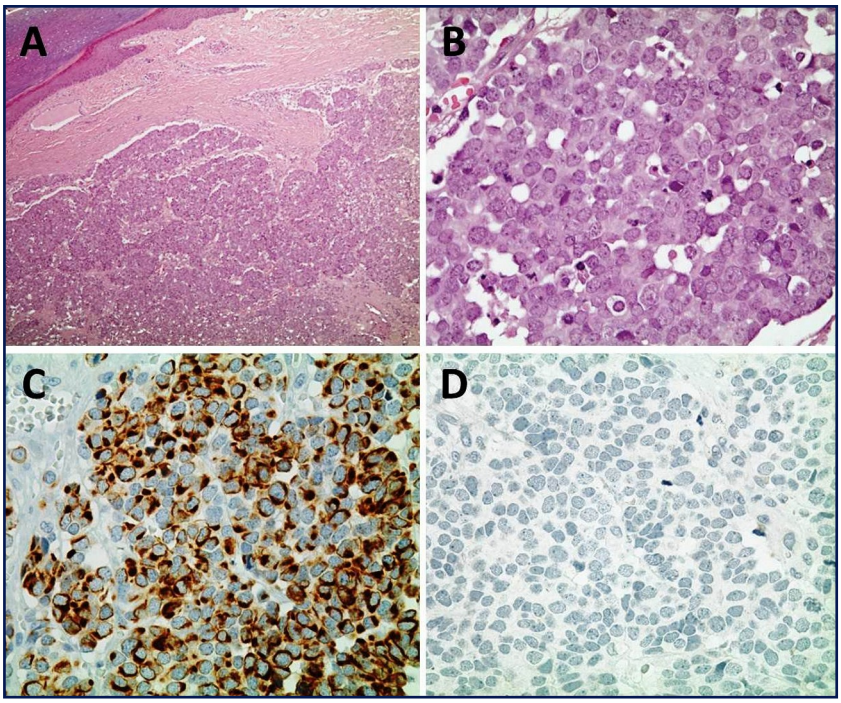


**Figure 2**

*A) Tumor grows with a solid pattern within the dermis (hematoxylin and eosin x 5);*
*B) Sheets of small round cells with hyperchromatic nuclei and scanty cytoplasm (hematoxylin and eosin x40);*
*C) Tumor cells are positive for cytokeratin 20 and show the characteristic immunohistochemical paranuclear dot-like staining pattern (cytokeratin 20 x 40);*
*D) Tumor cells are immunonegative for cytokeratin 7 (x 40).*

As Zalaudek *et al*.[11] stated, milky red areas are very uncommon in benign lesions (with the exception of pyogenic granuloma, dermoscopically undistinguishable from amelanotic melanoma), and this feature within a lesion has to be considered as an indication to the excision.

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