

Case for diagnosis Caso para diagnóstico

Bernardo Gontijo¹
Flávia Vasques Bittencourt³
Angélica Duarte Oliveira Rocha⁵

Thaíssa Oliveira de Almeida Coelho²
Glaysson Tassara Tavares⁴

CASE REPORT

An 80-year-old Caucasian male patient was referred for evaluation of a rapidly growing, asymptomatic, erythematous nodule of 2 cm in diameter on his left cheek (Figure 1A). The lesion had been present for four months and the patient was otherwise healthy. Dermoscopy revealed a homogeneous pink background with polymorphous telangiectatic vessels (linear, rounded and comma-shaped) (Figure 1B).

Histopathologic examination of the excision biopsy specimen showed tumoral masses extending

from the deep dermis to the subcutaneous fat tissue (Figure 2A). At greater magnification, tumor cells showed uniform nuclei and scant cytoplasm (Figure 2B). Immunohistochemical staining was positive for Cytokeratin 20 (CK20) in a paranuclear, dot-like pattern (Figure 3).

Ipsilateral cervical lymph node enlargement was detected on follow-up. Cytological analysis of the aspiration biopsy material showed atypical cells indicating a possible lymphatic extension of the disorder.

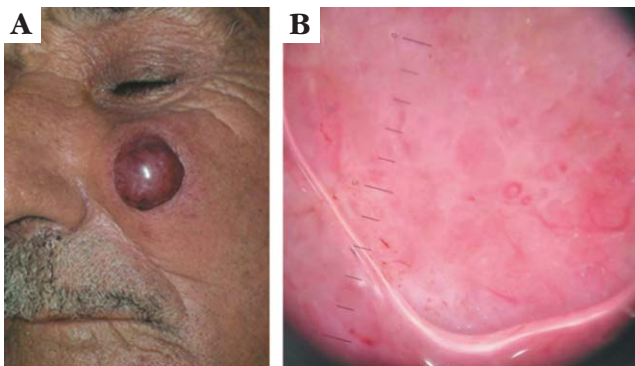


FIGURE 1: Erythematous nodule on the left cheek (A); Dermoscopy: pink background with linear, round and comma-shaped telangiectasias (B)

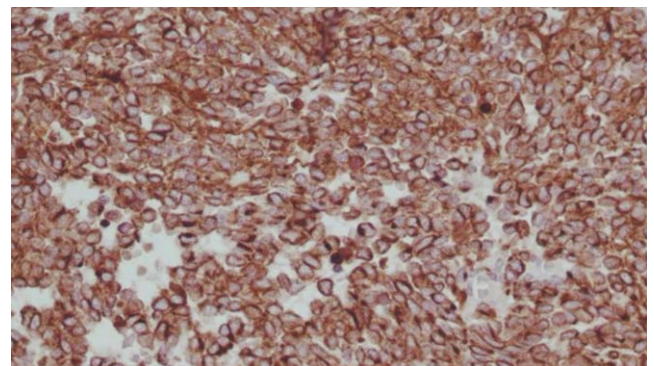


FIGURE 3: Paranuclear positive staining for CK20

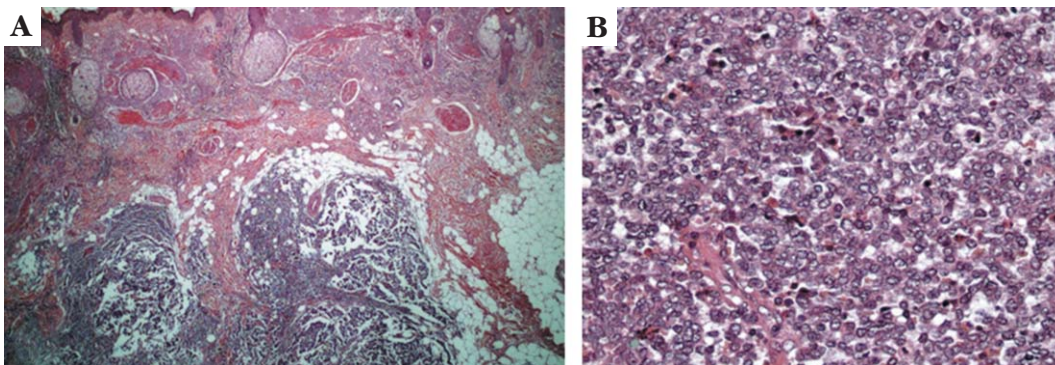


FIGURE 2: Tumoral masses in the deep dermis and subcutis (A); Round tumor cells with scant cytoplasm and uniform nuclei (B)

Received on 12.07.2011.

Approved by the Advisory Board and accepted for publication on 18.09.2011.

* This study was conducted at the Dermatology Unit, Teaching Hospital, Federal University of Minas Gerais - Belo Horizonte (MG), Brazil.

Conflict of interest: None

Financial funding: None

¹ MD, PhD. Associate Professor of Dermatology, Medical School, Federal University of Minas Gerais (UFMG) - Belo Horizonte (MG), Brazil.

² Medical Resident. Dermatology Unit, Teaching Hospital, Federal University of Minas Gerais (HC-UFGM) - Belo Horizonte (MG), Brazil.

³ MD, PhD. Adjunct Professor of Dermatology, Medical School, Federal University of Minas Gerais (UFMG) - Belo Horizonte (MG), Brazil.

⁴ Dermatologist. Dermatology Unit, Teaching Hospital, Federal University of Minas Gerais (HC-UFGM) - Belo Horizonte (MG), Brazil.

⁵ Dermatologist. Octaviano Neves Maternity Hospital - Belo Horizonte (MG), Brazil.

DISCUSSION

Merkel cell carcinoma (MCC) is an uncommon, aggressive and often lethal skin malignancy with neuroendocrine differentiation. It predominantly affects sun-exposed areas of elderly Caucasian and immunosuppressed patients.¹

Although a rare entity, MCC incidence rates showed a threefold increase between 1986 and 2001 in the United States.² This could be attributed to the aging of the population, the emergence of novel diagnostic tools and a growing number of immunosuppressed individuals.

In 2008, Feng et al. described a novel human polyomavirus (PyV) associated with MCC.³ Unlike other human PyV, those detected in MCC show oncogenic potential⁴. Furthermore, these PyV have been isolated from cutaneous swabs obtained from healthy adults and also from environmental surfaces such as door handles and vending machine keyboards.⁵ These findings suggest that Merkel cell PyV may cause a long-standing, asymptomatic infection in the skin.

The usual clinical picture in cases of MCC is that of a solitary, rapidly enlarging, non-tender nodule that is pink to violaceous in color. The head and neck are the most commonly affected areas followed by the extremities and buttocks. Due to the absence of speci-

fic or unique clinical features, MCC is rarely suspected at physical examination. Differential clinical diagnoses include basal and squamous cell carcinoma, amelanotic melanoma, pyogenic granuloma, keratoacanthoma and metastatic neoplasms.¹ Accurate diagnosis, therefore, relies on histopathologic and immunohistochemical findings. Reports of dermoscopy findings are rare and no conclusive standard pattern has been established so far.

Neoplastic Merkel cells are small and round, with scant cytoplasm and uniform nuclei (small, round, blue cell tumors), densely packed in nests, cords or sheets and located from the dermis to the subcutis. Normal Merkel cells show diffuse CK20 staining whereas malignant ones show a paranuclear dotted pattern.⁶

The 10-year survival rate is higher in women (64.8%) than in men (50.5%).⁷ The association with PyV has prognostic implications since patients with PyV-negative MCC are more likely to show regional nodal metastasis at diagnosis.⁸

Surgery is the mainstay of treatment and radiotherapy may be used either as monotherapy or adjuvant therapy. Since almost one-third of patients with clinically negative node involvement are found to have microscopic nodal disease, sentinel lymph node biopsy is recommended for all primary tumors.⁹ □

Abstract: An 80-year-old Caucasian male patient was referred for evaluation of a rapidly growing, asymptomatic, erythematous nodule measuring 2 cm in diameter on his left cheek. The lesion had been present for four months. Dermoscopy revealed a homogeneous pink background with polymorphous telangiectatic vessels. Histopathology showed tumors in the deep dermis and subcutis composed of round cells with scant cytoplasm. Immunohistochemical staining was positive for CK20 confirming the diagnosis of Merkel cell carcinoma.

Keywords: Dermoscopy; Immunohistochemistry; Merkel cell carcinoma; Neuroendocrine carcinoma

Resumo: Paciente de 80 anos, branco, sexo masculino, encaminhado para avaliação de nódulo eritematoso de 2 cm, assintomático, de crescimento rápido, localizado na região malar esquerda, com quatro meses de evolução. À dermatoscopia visualizou-se fundo homogêneo róseo com telangiectasias polimorfas. O exame anatomopatológico revelou massas tumorais atingindo a derme profunda e o subcutâneo, compostas por células arredondadas, de citoplasma escasso. A imunohistoquímica foi positiva para CK20, confirmando o diagnóstico de carcinoma de células de Merkel. **Palavras-chave:** Carcinoma de célula de Merkel; Carcinoma neuroendócrino; Dermatoscopia; Imunohistoquímica

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MAILING ADDRESS:

Bernardo Gontijo

**Rua Domingos Vieira, 300 suite 505 - Santa Efigênia
30150-240 Belo Horizonte, MG, Brazil**

Phone/Fax: 55-31-3241-1185 / 55-31-3241-6691

E-mail: bernardogontijo@terra.com.br

How to cite this article: Gontijo B, Coelho TOA, Bittencourt FV, Tavares GT, Rocha ADO. Case for diagnosis. Merkel cell carcinoma. *An Bras Dermatol.* 2012;87(3):491-2.