Merkel Cell Carcinoma: A Case Report and Review of Literature

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Abstract

Merkel cell carcinoma (MCC) is a highly aggressive tumor (WHO, 2018), primarily affecting elderly individuals with a history of chronic sun exposure. Researchers link factors such as ultraviolet radiation, immunosuppression, and infection with the Merkel cell polyomavirus (MCPyV) to its etiology. Despite ongoing debate over its cell origin, MCC's immunohistochemical profile and morphological characteristics resemble those of native Merkel cells found in the skin. We present a case observed in the pathology department at the Mohamed VI University Hospital Center in Marrakech. This case study involves a 79-year-old patient without any specific medical history who presented with a rapidly enlarging mass on the upper lip. Merkel cell carcinoma is a highly aggressive skin tumor associated with various causative factors, including UV radiation, immunosuppression, and MCPyV infection. Its diagnosis relies on careful histopathological examination and immunohistochemical profiling. Differential diagnosis considerations are crucial to ensure accurate classification and appropriate management of this challenging malignancy.

Keywords: *Merkel cell, Carcinoma, Pathology, Upper lip*

Introduction

Merkel cell carcinoma (MCC) is a highly aggressive tumor,[1] primarily affecting elderly individuals with a history of chronic sun exposure. Researchers link factors such as ultraviolet radiation, immunosuppression, and infection with the Merkel cell polyomavirus (MCPyV) to its etiology. Despite ongoing debate over its cell origin, MCC's immunohistochemical profile and morphological characteristics resemble those of native Merkel cells found in the skin.

Observation

We present a case observed in the pathology department at the Mohamed VI University Hospital Center in Marrakech. This case study involves a 79-year-old patient without any specific medical history who presented with a rapidly enlarging mass on the upper lip. A biopsy was performed, and histological examination revealed an infiltrating malignant tumor proliferation organized into nests, trabecular structures, and diffuse sheets. The tumor cells were of moderate size, often with vesicular nuclei displaying a "salt and pepper" chromatin pattern. Numerous mitotic figures and mitonecrosis were evident. The cytoplasm was scant and basophilic. The reactive stroma was highly vascularized and accompanied by lymphovascular invasion. Immunohistochemical analysis demonstrated perinuclear dot-like staining with anti-CK20 antibodies, positivity for chromogranin, and neurofilament. Anti-CD45 and Anti-PS100 antibodies yielded negative results. The clinical course was marked by rapid tumor growth, leading to extensive surgical excision with lymph node dissection **(Figure 1)**.

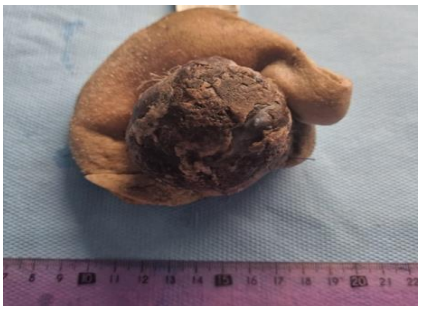

Figure 1. Grossig Specimen showing the mass on the upper lip.

Results and Discussion

Blue cells packed densely define Merkel cell carcinoma as a neuroendocrine carcinoma. The tumor predominantly arises in the dermis, often involving the overlying epidermis and occasionally invading subcutaneous fat. MCC exhibits a variety

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of growth patterns, including sheet-like formations, nests, and occasionally ribbons.[2, 3] Cell outlines may confluence or mimic lymphocytes, with frequent mitotic activity and potential necrosis. Lymphovascular invasion is a common feature. Furthermore, MCC may display uncommon squamous or sarcomatoid differentiation, and coexistence with actinic keratosis and in situ squamous cell carcinoma is frequently observed in the adjacent skin.[4]

To confirm the diagnosis of MCC and provide insights into its prognosis, immunohistochemical studies are indispensable.[5, 6] A typical feature is perinuclear staining with CK20, and positive stains include CD56, chromogranin, synaptophysin, and neurofilament **(Figure 2)**.[7]

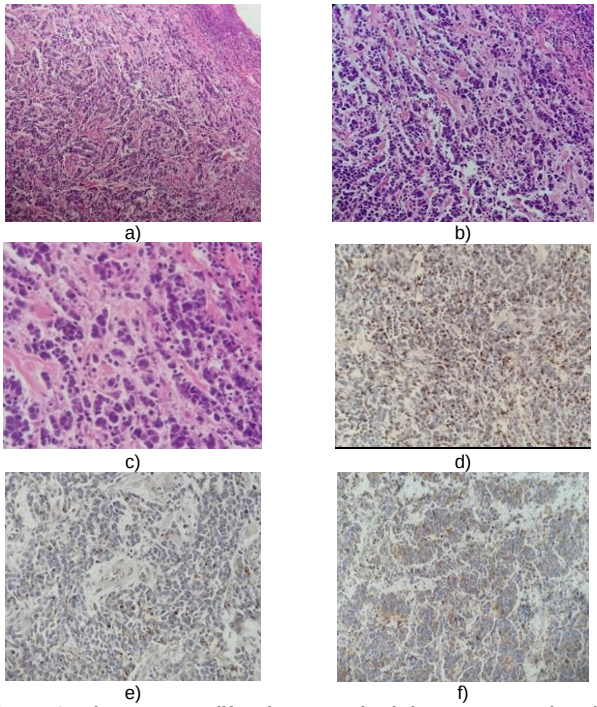

Figure 2. a) Tumor proliferation organized into nests, trabecular structures, and diffuse sheets. b) The tumor cells were of moderate size, often with vesicular nuclei displaying a "salt and pepper" chromatin pattern. c) Perinuclear dot-like staining with anti-CK20. d) and NF. e) Cytoplasmic staining for chromogranin.

Differential diagnosis considerations for Merkel cell carcinoma (MCC) encompass several possibilities: One potential differential diagnosis is Small Cell Lung Carcinoma, which may share morphological similarities with MCC.[8] However, a key distinguishing factor is the absence of CK20 staining and the presence of TTF-1 positivity, typically negative in MCC. Another challenging distinction is with Basal Cell Carcinoma. MCC can occasionally be confused with basal cell carcinoma due to the presence of densely packed blue cells. A meticulous examination of cellular features is essential, and when necessary, immunohistochemical studies can play a pivotal role in achieving accurate differentiation. Furthermore, distinguishing MCC from Melanoma and Squamous Cell Carcinoma demands a thorough assessment, incorporating scrutiny of tissue characteristics and a correlation with immunohistochemical findings to ensure a precise diagnosis.[9, 10]

Conclusion

Merkel cell carcinoma is a highly aggressive skin tumor associated with various causative factors, including UV radiation, immunosuppression, and MCPyV infection. Its diagnosis relies on careful histopathological examination and immunohistochemical profiling. Differential diagnosis considerations are crucial to ensure accurate classification and appropriate management of this challenging malignancy.

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Conflict of interest

None.

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Ethics statement

None.

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