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CASE REPORT

Merkel Cell Carcinoma on the Forearm: A Case Report and Literature Review

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Abstract: Merkel cell carcinoma (MCC) is a rare cutaneous growth with aggressive nature. It mostly affects on the head and neck of white men aged 65 years old and immunosuppressed patients. Limited cases were reported in Asian patients. Loco-regional metastases with the regional lymph nodes involvement were common. The treatment methods include complete surgical excision and radiotherapy. Topical imiquimod and biologics are promising therapies. Here, we present a case of MCC occurring in a 76-year-old Chinese woman.

Keywords: Merkel cell carcinoma, skin cancer, carcinoma

Merkel cell carcinoma (MCC) is a rare neuroendocrine skin malignancy with aggressive feature.[1] It mostly affects elderly white males above 65 years old and immunosuppressed patients with head and neck predilection.[2] Long time ultraviolet exposure and Merkel cell polyomavirus (MCV) are linked to the pathogenesis of MCC.[3] Here, we report a case of MCC occurring in a 76-year-old Chinese woman. Written informed consent for publication of the details was obtained from the patient.

Case Presentation

A 76-year-old Han Chinese previously healthy woman present to the clinic with an indurated, nontender, scarlet red, dome-shaped nodule on her right volar wrist (Figure 1). The growth enlarged rapidly in the last 6 months. Physical examination showed a red-to-violet subcutaneous nodule sized in the diameter of 2 cm. A biopsy was performed to confirm the diagnosis. On the standard hematoxylin-eosin stain, a dermal proliferation was noticed with nests of small blue basaloid cells and a "salt-and-pepper" chromatin pattern with scant cytoplasm and frequent mitotic nucleus (Figures 2 and 3). Cytokeratin-20 (CK20) immunohistochemical stain illustrates a typical paranuclear dot pattern (Figure 4). The tumor cells express synaptophysin (Syn) and epithelial membrane antigen (EMA). Also, CD99, S-100, lymphocyte common antigen (LCA) and thyroid transcription factor 1 (TTF-1) are negative, which is consistent with the diagnosis of MCC. The patient reported no lymphadenopathy and refused the sentinel lymph node biopsy. The tumor was removed with 1-cm margins and the defect repaired with an advancement flap. Positron emission tomography (PCT)/CT revealed no evidence of distant disease. At the patient's 6-month postoperative encounter, no signs of local recurrence or metastasis were noted. She will be followed closely every 6 months.

Discussion

MCC was first described by Cyril Toker in 1972 and named due to its tumor cells resembling Merkel cells, the mechanoreceptors for gentle touch which lie in the basal layer of the epidermis.[1] MCC is highly aggressive with approximately 30% mortality, which was higher than that observed in melanoma.[3] Loco-regional metastases with the regional lymph nodes involvement were common. The clinical presentation of MCC is varied. A solitary erythematous or

Received: 16 February 2022
Accepted: 24 May 2022
Published: 14 June 2022

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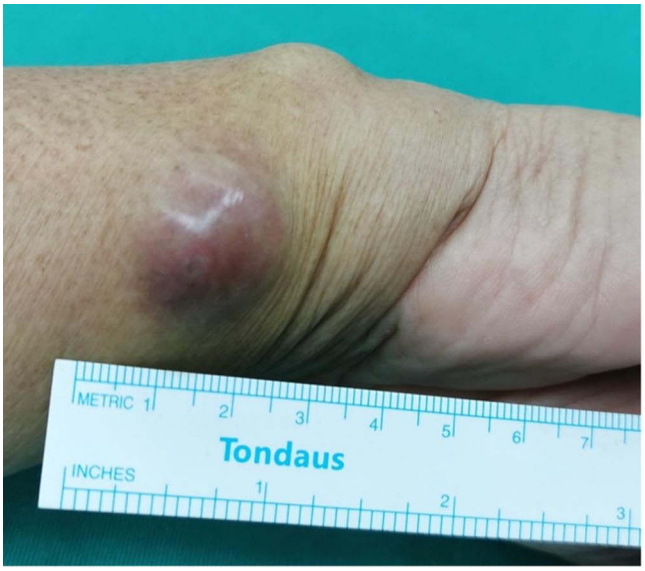

Figure 1 A red-to-violet subcutaneous nodule sized in the diameter of 2 cm on the right volar wrist.

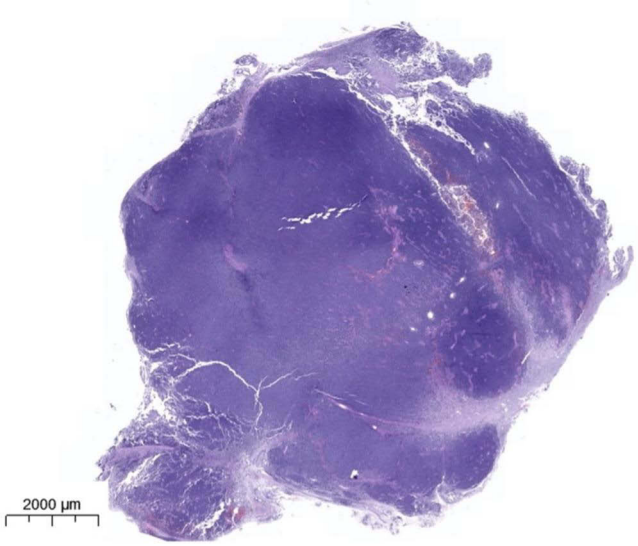

Figure 2 Hematoxylin-eosin stain shows large nodular collections crowded basaloid cells in the dermis.

violaceous nodule without ulceration on sun-exposed area especially on the head or neck in white men was noticed as the common lesion.[4] Other lesions such as subcutaneous masses, pediculated lesions, and telangiectatic papules are also reported.

MCC is 25 times more common in whites compared to others. Studies in Northern European showed MCC affects men more than women. However, epidemiologic studies in Africa and Asia are scant due to limited numbers of cases reported.[5] In this study, we reported a Chinese Han woman patient with no history of administration of immunosuppression may add evidence for the scarcely Asian MCC patients.

The pathogenesis of MCC is unclear; whether it is derived from epidermal Merkel cells, dermal neuroendocrine cells, or epidermal pluripotent stem cells is still a controversy. It has been reported to have disproportionately higher relevance to the long-term iatrogenic immunosuppression and lymphoproliferative disorders. The predominance of MCC in aged people indicated the important role of immunosenescence. Ultraviolet radiation also participated in the process of pathogenesis on MCC. Patients with psoriasis who received UVA therapy showed 100 times higher risk of developing

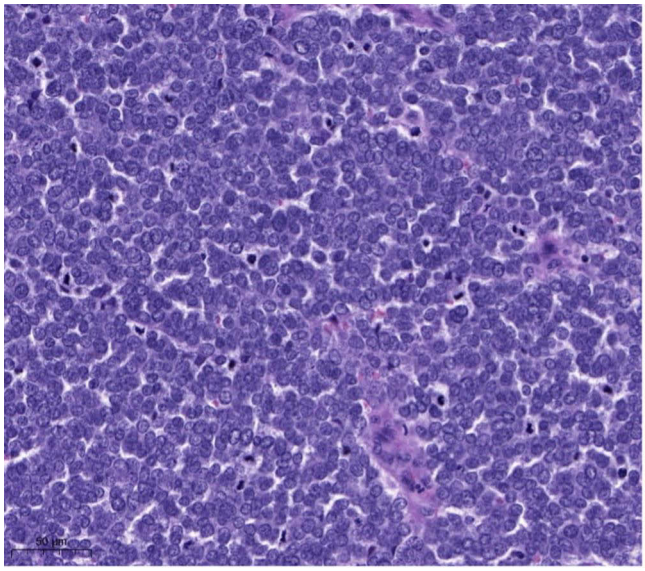

Figure 3 High-power magnification shows typical nuclear features with round nuclei with granular chromatin and scant cytoplasm. Scattered mitotic figures are present.

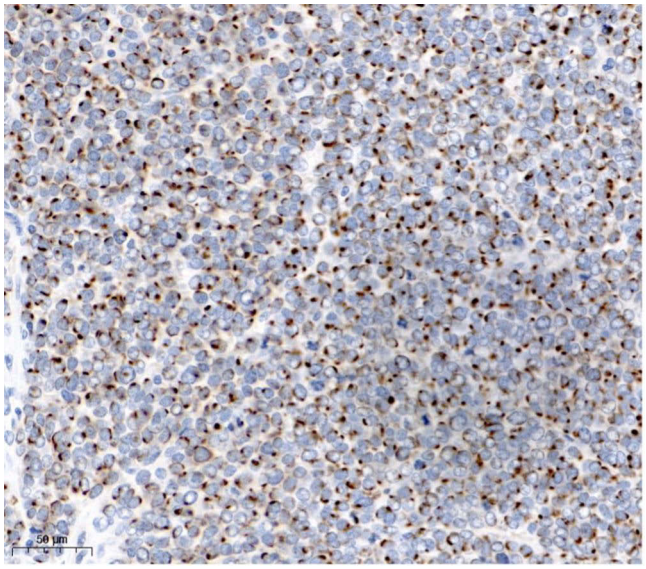

Figure 4 Cytokeratin-20 immunohistochemical staining shows a paranuclear dot pattern (original magnification: X200).

MCC than the general population. The discovery of MCV also provided insight into the pathogenesis of MCC. MCV was sequenced in 8 of 10 MCC tumors sampled, and viral DNA was integrated into the genome in 6 of 10 tumors.[6–8]

According to the National Comprehensive Cancer Network (NCCN) guideline for MCC diagnosis, a six-step algorithm was created. Clinical examination is always the first step in MCC diagnosis. A specimen biopsy of suspicious lesion was necessary. Histopathologically, tumors often originate in the dermis and often invade the subcutaneous fat layer. In 10% of patients, tumors spread to the epidermis, including the formation of Pautrier microabscesses. Therefore, it should be differentiated from skin T-cell lymphoma and superficial disseminated melanoma.

Immunohistochemical tumor cells express epithelial antigens. In addition to showing neuroendocrine characteristics, the most characteristic of skin neuroendocrine cancer is the expression of low molecular weight keratin (CAM5.2) and more specifically CK20. CK7 and TTF-1 are negative, which helps to distinguish primary skin neuroendocrine carcinoma

from skin metastases of bronchial small cell lung cancer. Tumor cells also often express nerve-specific enolase (NSE) and EMA but are negative for vimentin and S-100.[5,6]

As for treatment, complete surgical excision of the primary site is typically the first step. Current guidelines recommend 1–2cm margins down to fascia or periosteum. Radiotherapy is an alternative treatment for non-surgical cases.[8] Topical imiquimod is also a promising option. A case report chose 5% topical imiquimod cream, applied once daily for 5 days each week for a total period of 35 weeks, noticeable remission confirmed its efficacy.[9] Biologics were also introduced to the therapeutic regime. Avelumab, a human anti-programmed death-ligand 1 (PD-L1) monoclonal antibody, was administrated to 88 distal metastasis MCC patients for 12 months. A 72.4% of responses were noticed in that last 1 year, which suggests a promising treatment and a possible treatment target for MCC.[10]

Conclusion

MCC is a rare tumor especially in Chinese patients. Early diagnosis and ful surgical removal are important for clinicians. Regional lymph node examination is also necessary. This review may help dermatologists get more familiar to the clinical presentation and therapeutic developments of MCC.

Acknowledgments

Publication of the case details was approved by Beijing Chaoyang Hospital, Capital Medical University.

Disclosure

The authors report no conflicts of interest in this work.

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