Merkel cell carcinoma-derived Erysipelas carcinomatosum

Dear Editor,

Merkel cell carcinoma (MCC) is a malignant tumor of the skin, which affects elder individuals. Cumulative sun exposure, immunosuppression, and solid organ transplantation are risk factors for this anaplastic, highly aggressive neoplasia (Becker et al. 2017).

Carcinoma erysipeloides (CE) is an uncommon form of metastasis in which malignant cells spread to skin via superficial dermal lymphatic vessels. To date, there has been no report of CE due to MCC of the skin. Herein, we present the first case of CE due to MCC invasion of the skin (Krumbholz et al., 2006).

A 91-year-old male presented with a 6-month history of gradually progressive, asymptomatic, extensive lesions, which started from neck. Physical examination revealed a thick, reddish-purplish nodules and plaques involving the head & neck and trunk area. Lesion on the neck was thicker, ulcerated, and oozing (Figure 1). These features made us to perform a prompt biopsy and histopathology of the lesion showed an extensive infiltration of the entire dermis and epidermis with monotonously uniform, small, round to oval cells with a high mitotic rate (Figures 2 and 3). Immunohistochemistry stains were positive for p63, CK-20, synaptophysin and negative for thyroid transcription factor-1. With these findings, our patient has been diagnosed as CE due to MCC invasion. Chest CT demonstrated extensive infiltration of the cutis, and subcutis without any metastasis. Patient was immunocompromised due to myeloproliferative neoplasia and under treatment of ruxolitinib. Considering the lesion’ location and the size, avelumab therapy initiated. At 3-month follow-up, a partial response was observed.

MCC is an uncommon but highly aggressive cutaneous cancer with neuroendocrine characteristics. It commonly affects the skin of head and neck and is majorly seen in fair-skinned elderly population. MCC risk is significantly increased in patients with other malignancies (multiple myeloma, chronic lymphocytic leukemia, non-Hodgkin lymphoma, etc.) (Emge & Cardones, 2019; Safa et al. 2018). Merkel cell
polyomavirus (MCPyV) is another risk factor to develop MCC. Eighty percent of the individuals are infected by MCPyV. MCC is regarded as the most lethal cutaneous cancer and the incidence and mortality of this highly metastatic tumor is increasing (Czech-Sioli et al. 2020). There have been substantial progression recently in understanding of etiology, diagnosis, and management of MCC; however, much about its true history and features remains unclear. Most cases of CE are due to underlying adenocarcinoma, mostly breast carcinomas. However, other adenocarcinomas such as thyroid, parotid gland, larynx, prostate, pancreas, stomach carcinomas, and even melanoma have been reported as a cause. CE rarely can be the first sing of the tumor itself, but it usually appears after chemo-radio therapy of surgery. It is thought that these treatments may lead shedding of the cells through lymphatics (Vandersee, 2019). In our patient, tumor occurred de novo without any intervention and progressively expanded.

In conclusion, this case represents a case of CE due to MCC of the skin. When clinicians notice rapidly progressive, erythematous plaques or nodules on an elderly immunocompromised individual, MCC should be kept in mind to diagnose early and treat immediately this highly mortal tumor.

CONFLICT OF INTEREST
The authors declare no potential conflict of interest.

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