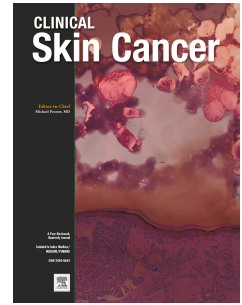


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Merkel Cell Carcinoma: Characteristics, Management, and What's on the Horizon.

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Conflicts of interest: None

Abstract: Merkel cell carcinoma is a rare and aggressive cutaneous malignancy with a steadily rising incidence. Advancing age, fair skin, ultraviolet light exposure, and immunocompromise are risk factors. Sixty to eighty percent are associated with the Merkel cell polyomavirus, and the remainder carry a high mutational burden. Recently immunotherapy has been shown to be a highly effective and well tolerated treatment. The disease is also very chemosensitive similar to small cell lung cancer. Ongoing trials are evaluating the role of adjuvant immunotherapy and novel immunotherapy agents as well as targeted therapy and endocrine therapy with somatostatin analogues. Also included in this review is our institution's experience with MCC in 6 patients treated with pembrolizumab, 5 of whom experienced an excellent response.

Key words: Merkel cell carcinoma, trabecular carcinoma, primary small cell carcinoma of the skin, incidence, Merkel Cell polyomavirus, clinical trials, pazopanib, imatinib, checkpoint inhibitors, chemotherapy, octreotide.

Introduction

Merkel cell carcinoma (MCC) is a rare cutaneous malignancy whose incidence is rising. It is also called "primary small cell carcinoma of the skin" due to its neuroendocrine features and aggressive clinical course. This cancer presents a diagnostic challenge given both its low incidence and relatively innocuous appearance. Generally MCC begins as a rapidly expanding but asymptomatic purplish to red colored smooth papule/plaque with irregular borders usually in sun exposed area in fair-skinned individuals over 50. The tumor cells are usually positive for CK20 [1].

Epidemiology: As of 2013 MCC occurs with a frequency of about 0.7 per 100,000 age adjusted person-years in the United States [2]. The SEER 18 database, which covers 28% of the United States population, recorded 654 cases in 2013 (Figure 1a) and the American Cancer Society estimates the total number of cases in the United States to be around 1500 for the year 2017 [3]. This is in comparison to melanoma with an incidence of 23.3 cases per 100,000 in 2013, which in turn represented 5.39% of all new cancer cases [4]. MCC predominantly affects fair-skinned individuals with 95% of MCCs occurring in Caucasians [5]. There is a male predominance with twice as many men being affected as women. Frequency also rises with advancing age, with a mean age of diagnosis of 75 and peak incidence

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