

CASE STUDY

Merkel Cell Carcinoma

Ardalan Minokadeh, PhD; Adam J. Wulkan, BS; Kenneth Beer, MD; Jill S. Waibel, MD

 AU: Please
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 Author
 Affiliations

A 92-year-old man presented for evaluation with a 1-month history of a rapidly growing asymptomatic pink nodule on his forearm. Biopsy results of the lesion demonstrated pathology consistent with Merkel cell carcinoma (MCC). Immunohistochemical studies displayed positive cytoplasmic staining for cytokeratin AE1/AE3, positive dot-like perinuclear staining for cytokeratin-20, diffuse cytoplasmic staining for neuron-specific enolase, and no significant staining for S-100. Subsequent positron emission tomography (PET) did not reveal evidence of metastatic disease. Wide excision of the lesion was performed along with a sentinel node biopsy of his left axilla. The sentinel nodes were negative for MCC. Adjuvant radiation treatment of the tumor site was provided because the pathologist noted MCC within 2 mm of the deep margin.

Six months after his first nodule was discovered, the patient presented with a 1-week history of an 8-mm pink papule on his right preauricular area (Figure 1). A shave biopsy was performed, which revealed MCC (Figure 2 and Figure 3). Immunohistochemical studies revealed a strong expression for cytokeratin-20 within the atypical cells, and there was no evidence of thyroid transcription factor-1 expression. Results from repeat PET were negative for metastatic disease. The patient underwent a wide local excision of this second tumor with reconstruction, without complications. Subsequently, he completed a course of adjuvant radiation therapy. In the 6 months after that surgery, the patient had no recurrences, metastases, or new lesions in the sites of either of his cancers. Although it has been presumed that these are two primary MCCs, this cannot be verified.

DISCUSSION

MCC is a rare, aggressive neuroendocrine tumor arising from Merkel cells, which are specialized neuroreceptors located in the basal layer of the epidermis. A 33% mortality rate is associated with this diagnosis, higher than that of melanoma.¹ There are about 1500 new cases of MCC in the United States each year, and the incidence of this cancer is on the rise.² Recent studies in the etiology of this malignancy have identified that the Merkel cell polyomavirus (MCV) may be responsible for inducing MCC.³ DNA sequences of the MCV have been detected in 80% of tumors compared with only 8% to 16% of controls. In the following case discussion, we present a patient with two MCC lesions each lacking the MCV, with a discussion of the etiology and treatment of this malignancy.

At present, it is believed that many MCCs have a viral etiology. The identification of the MCV at the University of Pittsburgh has renewed interest in MCC.³ Initial studies have demonstrated a monoclonal pattern of DNA integration into MCCs, suggesting that the viral integration occurs prior to clonal tumor growth. This supports the idea that the virus is responsible for MCC, as opposed to MCCs merely having increased susceptibility to be

secondarily infected by MCV.⁴ In a recent review of MCC, the incidence of MCC is estimated to be 0.6 per 100,000,⁵ representing a rise from historic incidence rates. The need for novel therapeutic interventions is clear. Indeed, a potential outcome of this testing will be the development of antiviral therapy for MCV-positive patients, significantly altering the therapeutic landscape for MCC. Histochemical staining of the two tumors in this patient failed to demonstrate viral protein (Figure 3). Thus, it is likely that our patient is one of the approximately 15% to 25% of MCC patients who are MCV negative. One other possibility is that the patient in this report had infection with MCV that was not detected with present methodologies.

Although wide excision remains the mainstay of treatment for MCC, several studies have shown superior results utilizing Mohs surgery.^{6,7} Mohs surgery with adjunctive radiation has been shown to decrease recurrence rates. Sentinel node mapping in patients with MCC has been advocated by the new National Comprehensive Cancer Network guidelines for the management of patients with MCC; this modality has utility in monitoring MCC patients to determine the likelihood of distant metastasis.

Address for Correspondence: Jill Waibel, MD, 7800 SW 87th Street, Suite B200, Miami, FL 33173

 AU: Please
 Provide
 E-mail ID
 for cor-
 responding
 author.