



Metastatic Merkel Cell Carcinoma

Andrew Krane, MD¹, Vijairam Selvaraj, MD, MPH¹, Andrew Hsu, MD²

¹ Department of Medicine, Warren Alpert Medical School at Brown University,

² Division of Hematology Oncology, Department of Medicine, Warren Alpert Medical School of Brown University

Journal of Brown Hospital Medicine

Vol. 1, Issue 2, 2022

Article Information

Keywords: merkel cell carcinoma, elderly, uv radiation, skin cancer, melanoma, basal cell carcinoma, squamous cell carcinoma, metastatic
<https://doi.org/10.56305/001c.34649>

Submitted: April 15, 2022 EST

Accepted: April 22, 2022 EST

Abstract

Merkel cell carcinoma is a rare, aggressive skin cancer that affects mainly the sun exposed areas in the elderly, especially the head and neck. Immunohistochemical analysis is necessary to establish diagnosis. We describe an elderly male who presented with hematochezia and was incidentally noted to have liver lesion on abdominal imaging, which was subsequently diagnosed as Merkel cell carcinoma.

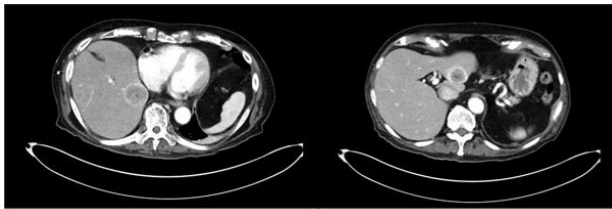


Figure 1. CT scan of abdomen pelvis showing hepatic lesions, along with portocaval adenopathy

An 86-year male with a history of Parkinson's disease, atrial fibrillation on apixaban, prostate cancer under surveillance, and Merkel cell carcinoma (MCC) of the scalp presented to the hospital with hematochezia. His hematochezia was managed in conjunction with gastroenterology and resolved spontaneously. A computerized tomography scan of his abdomen and pelvis obtained as part of his initial evaluation revealed multiple hepatic lesions and portocaval adenopathy (Figure 1). A liver biopsy was consistent with Merkel cell carcinoma. Furthermore, magnetic resonance imaging of the brain revealed an enhancing right occipital lobe lesion (Figure 2). Of note, his Merkel cell carcinoma was managed by wide excision 1.5 years prior to presentation with pathology noting lymphovascular space involvement and a positive margin. He completed a course of adjuvant radiation three months after his excisional surgery. Given evidence of metastatic disease along with his co-morbidities and poor performance status, the decision was made to focus on comfort-directed care, and he was transferred to an inpatient hospice facility.

Named after Friedrich S. Merkel, MCC was first described in 1972 by Cyril Toker when he reported five cases of 'trabecular carcinoma of the skin'.¹ Merkel cells are neuroendocrine cells along the basal layer of the epidermis. These cells proliferate and metastasize to the

lungs, brain, bones, and other organs. The incidence of MCC is 0.24 cases/100,000 persons.² Risk factors include light skin color, age, male sex, immunosuppression, and other malignancies.³ Pathogenic factors include ultraviolet radiation (UV) to sun-exposed areas, Merkel cell polyomavirus (dsDNA), and TP53 mutations. Typical locations of lesions include: head and neck (43%), upper limbs and shoulders (24%), lower limbs and hip (15%), trunk (11%), and other areas (9%).⁴ Clinical appearance is often non-specific, and usually presents as painless erythematous intradermal lesions, with a tendency to ulcerate as it grows bigger. Significant clinical features of this condition have been summarized with the acronym AEIOU (Asymptomatic, Expanding rapidly, Immune suppression, Older than 50yo, UV radiation).⁵ Differential diagnoses should include melanoma, basal cell carcinoma, lymphoma, metastatic neuroblastoma, and squamous cell carcinoma. The presentation may be local (65%), with regional lymph node metastasis (27%), or with distant metastases (8%). A definitive diagnosis requires a biopsy with cytokeratin 20 (CK-20) being a sensitive and specific marker of MCC. Management involves surgical resection with lymph node excision, radiation therapy, chemotherapy, and/or immunotherapy, depending on the stage.⁶

1. Stage I & II (<2cm vs >2cm or w/ invasion into bone/muscle/fascia/cartilage).
 - Surgery followed by radiation.
2. Stage III (transit metastasis or regional lymph node disease)
 - Surgery followed by radiation, Radiation therapy, Immunotherapy, Chemotherapy.
3. Stage IV (metastasis beyond the regional lymph nodes)

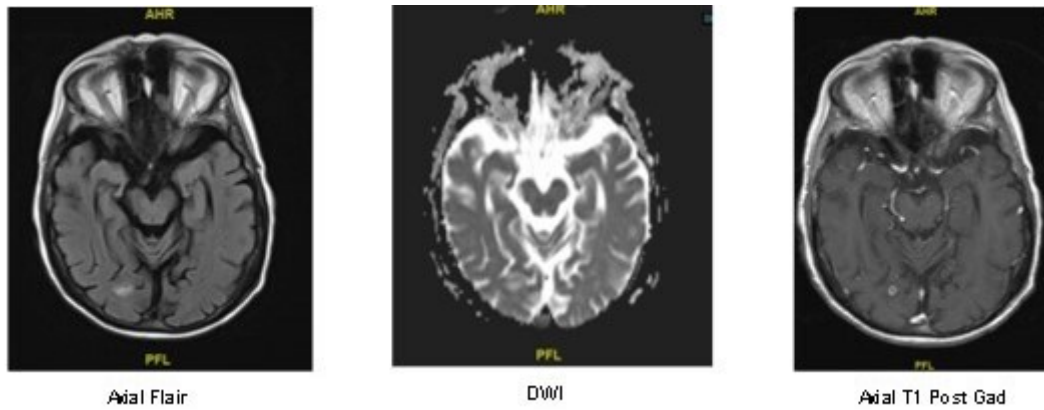


Figure 2. MRI brain images showing 0.6cm enhancing right occipital lobe lesion, suspicious for metastatic deposit.

- Immunotherapy (Pembrolizumab or Avelumab)
- Chemotherapy, surgery, or radiation therapy as palliative treatment to relieve symptoms and improve quality of life

In conclusion, physicians must have a high index of suspicion of MCC in the elderly population, especially in those patients with non-tender lesions and AEIOU clinical features. Given correlation between staging and survival, early diagnosis is essential to initiate early treatment to relieve symptoms and improve quality of life.

Corresponding Author

Vijairam Selvaraj, MD
 Assistant Professor of Medicine, Clinical Educator
 Warren Alpert Medical School at Brown University
 Division of Hospital Medicine
 The Miriam Hospital, 164 Summit Avenue, Providence,
 RI 02906

Conflicts of Interest

The authors declare they have no conflicts of interest.



This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CCBY-NC-4.0). View this license's legal deed at <https://creativecommons.org/licenses/by-nc/4.0> and legal code at <https://creativecommons.org/licenses/by-nc/4.0/legalcode> for more information.

REFERENCES

1. Toker C. Trabecular carcinoma of the skin. *Arch Dermatol*. 1972;105(1):107-110. [doi:10.1001/archderm.105.1.107](https://doi.org/10.1001/archderm.105.1.107)
2. Agelli M, Clegg LX. Epidemiology of primary Merkel cell carcinoma in the United States. *J Am Acad Dermatol*. 2003;49(5):832-841. [doi:10.1016/s0190-9622\(03\)02108-x](https://doi.org/10.1016/s0190-9622(03)02108-x)
3. Gass JK, Chan SK, Rytina E, Greenberg DC, Burrows NP. Multiple primary malignancies in patients with Merkel cell carcinoma. *J Eur Acad Dermatol Venerol*. 2010;24(5):601-603. [doi:10.1111/j.1468-3083.2009.03458](https://doi.org/10.1111/j.1468-3083.2009.03458)
4. Harms KL, Healy MA, Nghiem P, et al. Analysis of Prognostic Factors from 9387 Merkel Cell Carcinoma Cases Forms the Basis for the New 8th Edition AJCC Staging System. *Ann Surg Oncol*. 2016;23(11):3564-3571. [doi:10.1245/s10434-016-5266-4](https://doi.org/10.1245/s10434-016-5266-4)
5. Heath M, Jaimes N, Lemos B, et al. Clinical characteristics of Merkel cell carcinoma at diagnosis in 195 patients: the AEIOU features. *J Am Acad Dermatol*. 2008;58(3):375-381. [doi:10.1016/j.jaad.2007.11.020](https://doi.org/10.1016/j.jaad.2007.11.020)
6. Tello TL, Coggs K, Yom SS, Yu SS. Merkel cell carcinoma: an update and review: current and future therapy. *J Am Acad Dermatol*. 2018;78(3):445-454. [doi:10.1016/j.jaad.2017.12.004](https://doi.org/10.1016/j.jaad.2017.12.004)