BRIEF ARTICLES

Merkel Cell Carcinoma Presenting as an Indurated Plaque

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ABSTRACT

Merkel Cell Carcinoma (MCC) is a rare and aggressive neuroendocrine carcinoma that most commonly occurs in elderly Caucasian males who have a history of extensive sun exposure. Most cases of Merkel Cell Carcinoma (MCC) present in the outpatient setting and include a single, asymptomatic, red or pink lesion on sun exposed areas with a diameter less than 2 cm. There was a threefold increase in the incidence of MCC from 1986 to 2001, and the incidence of MCC is predicted to continue to increase secondary to the aging “baby boomer” population. To our knowledge, there are only two case reports that describe MCC resembling and receiving the diagnosis of cellulitis at initial presentation. We report an unusual case of MCC presenting as suspected cellulitis of the chest wall in an elderly Caucasian male.

INTRODUCTION

Merkel Cell Carcinoma (MCC) is a rare and aggressive neuroendocrine carcinoma that most commonly occurs in elderly Caucasian men who have a history of extensive sun exposure. The incidence of MCC increased threefold from 1986 to 2001 and is predicted to continue to increase since the “baby boomer” population is aging, and the risk of MCC increases with age.[1,2] Most cases of MCC present in the outpatient setting and include a single, asymptomatic, red to pink lesion on sun exposed areas (most commonly in the head and neck region) with a diameter less than 2 cm.[3] However, diagnosis is often challenging due to a combination of low suspicion by clinicians and a range of possible clinical presentations. We report an unusual presentation of MCC in form of an indurated plaque with metastasis.

CASE PRESENTATION

A 93-year-old Caucasian man with a history of vitamin D deficiency, actinic keratoses, and basal cell carcinoma of the head and neck presented to the hospital with a 2-month history of progressive swelling, oozing, and redness of his right chest wall and right arm. He denied any associated chest pain. Dermatology was consulted for chest wall rash that was initially diagnosed as cellulitis by the primary team. Physical examination revealed an elderly cachectic male with an ill-defined erythematous, indurated, and edematous plaque involving most of right chest wall and extending to the right upper arm (Figure 1). The periareolar skin had a Peau d’orange appearance with a wound draining serosanguinous fluid. Right axillary, right supraclavicular, and left axillary lymphadenopathy was present.

Figure 1. Initial clinical presentation of MCC on right chest wall.

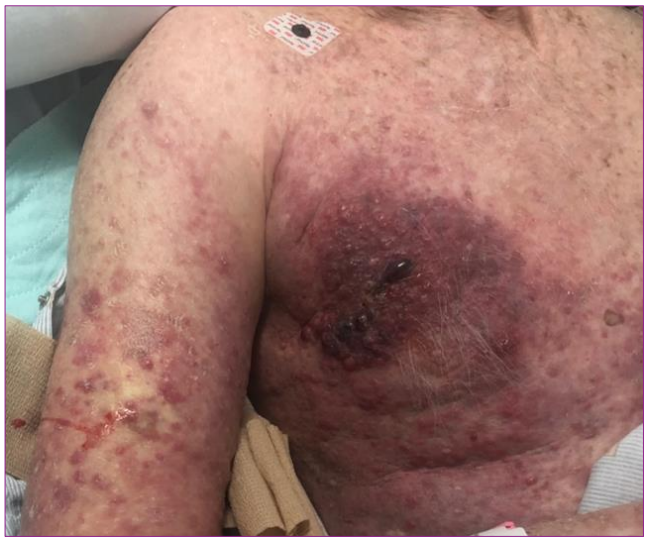
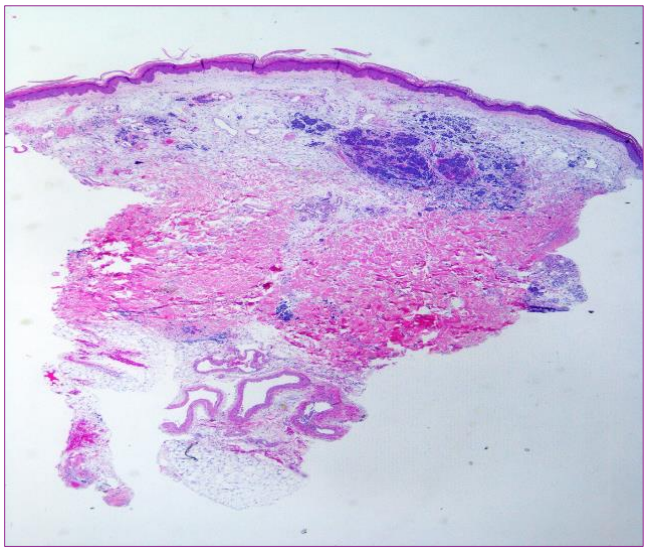
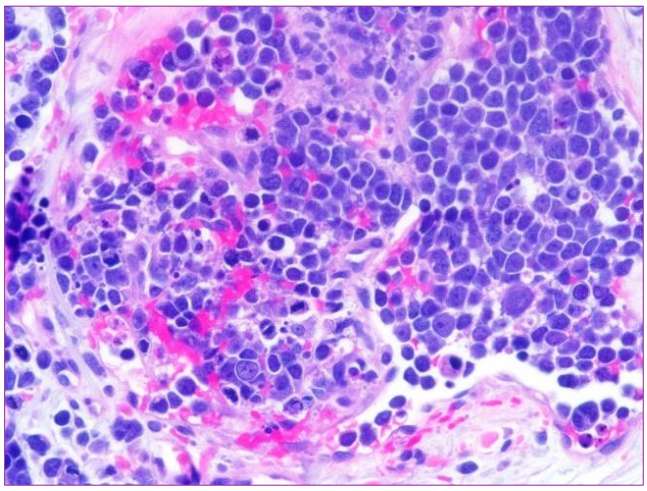


Figure 2. Histopathology of biopsy from right chest wall demonstrates dermal infiltrate of MCC.



A chest CT scan demonstrated a right sided pleural effusion, diffuse anasarca, bilateral lung opacities, diffuse adenopathy, diffuse skin thickening, and nodular infiltration of the right chest with possible involvement of the musculature. Biopsy of the chest wall exhibited a diffuse dermal infiltrate of polygonal cells with variably sized nuclei and finely granular "blastic" chromatin with occasional macronucleoli (Figures 2 and 3). The tumor cells showed paranuclear punctate positivity for CK20, CK8, panCK, synaptophysin, and neurofilament, while S100 and TTF-1 were negative. The diagnosis of MCC was made. Following discussion with his family, the patient decided to forgo treatment due to his age and extent of disease, choosing quality of life over quantity. He was discharged to hospice care and subsequently died.

Figure 3. Closer inspection of biopsy reveals polygonal cells with variably sized nuclei consistent with MCC.



DISCUSSION

Despite its increasing incidence, MCC is rarely suspected by clinicians prior to biopsy. In a review of initial clinical impressions of 106 MCC cases, the correct clinical diagnosis of MCC was made in only 1% of patients, with 56% of cases receiving benign clinical impressions initially.[3] Heath et al. created an acronym to guide clinicians in identifying common characteristics: AEIOU - Asymptomatic/lack of tenderness (88%), Expanding rapidly within 3 months (63%), Immune suppression (7.8%), Older than age 50 (90%), UV-exposed site (81%). In a

cohort study of 195 patients diagnosed with MCC from 1980 to 2007, 89% of patients met 3 or more of the AEIOU criteria.[3] Our patient met 3 of the criteria, as he had an asymptomatic/nontender lesion that expanded rapidly within 3 months and was older than age 50. However, the clinical appearance of our patient's tumor was atypical as the lesion was a large plaque as opposed to the typical 1-2 cm solitary lesion. Moreover, our patient was not immunocompromised, and the location of our patient's lesion was particularly rare, as Heath et al. observed trunk lesions at initial presentation in only 8% of patients.

While the use of AEIOU criteria is beneficial in terms of bringing awareness to typical MCC clinical phenotypes, it is important to know that clinical characteristics of MCC vary greatly and are nonspecific. There are recorded cases of MCC presenting with the clinical appearance of chalazion and cellulitis.[4,5] Other cases have had unusual morphologies like ulcerated and pedunculated masses.[4,6] Atypical age and location are also possible with reports of MCC occurring in young adults and teenagers and in the gluteal region and oral cavity.[4,7] As a result of the diverse and nonspecific clinical characteristics of MCC, definitive diagnosis is based on histopathologic means, with Cam 5.2, CK20 (characteristically paranuclear and dot-like), and neurofilaments being specific for MCC.[8,9]

MCC is an aggressive tumor with a propensity for nodal and distant metastasis, high rates of recurrence, and mortality rates that are higher than melanoma.[10] In an analysis of over 9,000 MCC cases from 1998-2012, lymph node involvement at initial presentation of MCC was documented in 26% of cases, with metastatic disease occurring in 8% of cases.[11] Extent of disease at presentation serves as a predictor of mortality, as local, nodal, and metastatic disease at presentation correlate to 51%, 35%, and 14% 5-year survival rates.[11] Patients who present with distant metastasis have a median survival time of 9 months, which is longer than the time our patient survived following his diagnosis.[12] The progressively poorer prognoses associated with more advanced cases of MCC emphasize the importance of early identification of at-risk patients with skin lesions suspicious for MCC.

Management of MCC varies based on disease stage and patient characteristics. A multidisciplinary approach in assessment and treatment is recommended as definitive therapeutic guidelines are not clearly established. Evaluation of regional and distant metastasis is required, with the dichotomy of clinically positive versus clinically negative lymph nodes serving as the first step in assessment.[13] Wide local excision of the primary lesion is typically recommended as the first step in treatment followed by adjuvant radiation therapy.[13] In metastatic cases, immunotherapy is now the preferred treatment modality as there appears to be an improved durability of response with immunotherapy compared to chemotherapy.[14] Clinical trials are currently evaluating avelumab, an anti-programmed death ligand 1 (PD-L1) agent, and pembrolizumab and nivolumab, anti-programmed cell death-1 (PD-1) receptor agents, with promising results.[15,16,17]

CONCLUSION

Our case covers an interesting and significant dermatologic topic as it describes a unique presentation of a rare tumor with a poor prognosis that is increasing in incidence and has emerging treatments.

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