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Case Report

A Rare Case of Merkel Cell Carcinoma of the Eyelid

Subtitle: Merkel Cell Carcinoma of the Eyelid: Diagnostic Insights and Management Strategies

Ingrid Raponi[1], MD, PhD, Angela Palma[2]*, MD, Pierluigi Alò[3], MD, Alessandra Perfetti[3], Caterina Tripodi[4] MD, PhD and Andrea Marzetti[4], MD

[1]Department of Maxillo Facial Surgery, F. Spaziani Hospital, ASL Frosinone, Italy

[2]Oral and Maxillo Facial Department Sapienza University of Rome, Italy

[3]Department of Pathology ASL Frosinone, Italy

[4]ENT and Maxillo Facial Department ASL Frosinone, Italy

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***Corresponding author:** Angela Palma, MD, oral and Maxillo Facial Department Sapienza University of Rome, Italy, Email: angela.palma@uniroma.it

ABSTRACT

Merkel cell carcinoma (MCC) is a rare and aggressive neuroendocrine malignancy of the skin, with a higher mortality rate than melanoma. This case report describes an 87-year-old woman presenting with a violaceous, nodular, exophytic mass on the upper eyelid. Histopathological examination revealed a poorly differentiated neoplasm with characteristic "salt-and-pepper" chromatin and a mitotic index of seven mitotic figures per eight high-power fields. Immunohistochemical analysis showed positive staining for CK20, Chromogranin and Synaptophysin, confirming the diagnosis of Merkel cell carcinoma. The tumor was staged as T2N0M0, with no evidence of lymph node or distant metastasis. This report highlights the diagnostic and management challenges associated with MCC, particularly in the eyelid, an uncommon but critical site. Early detection, histopathological evaluation and comprehensive staging, including sentinel lymph node biopsy, are essential for optimizing outcomes. Adjuvant radiotherapy and careful follow-up are recommended to minimize recurrence and address metastatic risk. This case emphasizes the importance of multidisciplinary care in managing this rare yet aggressive cancer.

Keywords: Merkel cell carcinoma; Eyelid; Surgery

Introduction

Merkel cells were first identified by Friedrich Merkel in 1875[1] and are in the basal layer of the epidermis, near the dermo-epidermal junction and in certain mucosal tissues, excluding the conjunctiva. Initially believed to function as mechanoreceptors (tactile sensory cells), these neuroendocrine cells are now thought to originate from the neuroectoderm and migrate to the skin alongside peripheral nerves. In 1978, Tang and Toker described the malignancy of these cells, which became known as Merkel cell carcinoma[2]. This cancer typically begins in the dermis, sparing the epidermal layer above it. Despite its neuroendocrine nature, Merkel cell carcinoma is not associated with hormonal syndromes[3].

Case Report

This study reports the case of an 87-year-old woman admitted to the ENT and Maxillo Facial Department of Spaziani Hospital, Frosinone, Italy unit with a violaceous mass on her upper eyelid **(Figure 1)**. During her hospitalization, an excisional biopsy was performed, ensuring clear margins. Local hemostasis was performed with absorbable hemostat of oxidized regenerated cellulose with neutral Ph (Oxitamp Powder®, Assut Europe) followed by a primary intention closure of the wound with non-absorbable suture (Assuplus®, Assut Europe). The tumor, described as a nodular, exophytic, solid and reddish mass, showed no signs of lymph node involvement or distant metastasis during the initial evaluation. Based on the American Joint Committee on Cancer (AJCC) classification system, the tumor was staged as T2N0M0 after the oncological surgery.

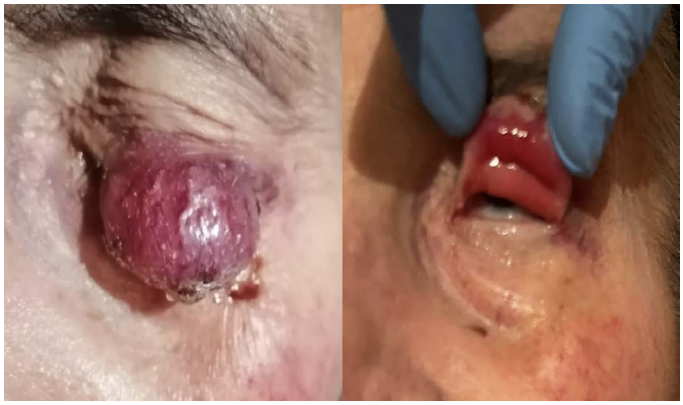

Figure 1: A clinical image shows a patient with Merkel cell carcinoma affecting the upper right eyelid. The tumor presents as a nodular, exophytic, solid and reddish lesion, causing mechanical ptosis due to its size and location.

The diagnosis was confirmed through histopathological evaluation, supported by immunohistochemical testing:

the histological analysis of a 2.5 cm skin biopsy, stained with hematoxylin and eosin, revealed an ulcerated epidermis. The superficial and deep dermis were extensively infiltrated by poorly differentiated neoplastic cells arranged in a solid growth pattern.

Cytological evaluation showed these cells had scant cytoplasm and round, well-defined, polymorphic nuclei displaying a characteristic “salt-and-pepper” chromatin pattern, with inconspicuous nucleoli. At 40x magnification, a mitotic index of seven mitotic figures per eight high-power fields was observed, along with scarce apoptotic cells. The tumor stroma contained minimal connective tissue strands and few vessels, some of which exhibited neoplastic embolization. No areas of necrosis were identified.

Immunohistochemical staining demonstrated positive expression for CK20, Chromogranin and Synaptophysin, while markers such as CDX2, S100 and Pan-Leu were negative. Based on the morphological and immunohistochemical findings, the diagnosis was consistent with Merkel Cell Carcinoma of the skin. Notably, there was no involvement of the epidermis **(Figure 2)**.

Despite the success of the procedure, further follow-up could not be carried out, as the patient unfortunately passed away due to cardiac complications unrelated to the cancer.

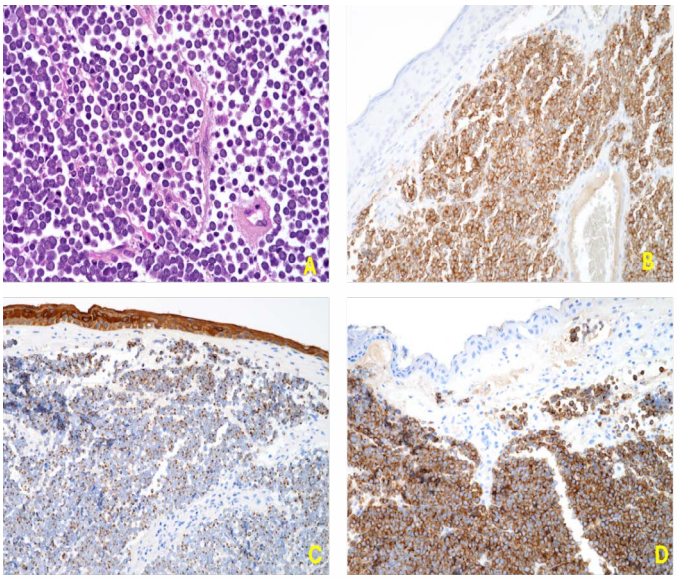

Figure 2: A: Hematoxylin-Eosin (H&E) Staining (40x); **B:** Pan cytokeratin Immunostaining (20x); **C:** Synaptophysin Immunostaining (20x); **D:** Chromogranin Immunostaining (20x).

Discussion

Merkel cell carcinoma is a rare and aggressive form of skin cancer, with a mortality rate surpassing that of melanoma (46% vs. 15% over five years). In this case report, the patient’s death was not directly linked to the carcinoma. Studies from the U.S. indicate an increase in incidence from 0.15 to 0.6 per 100,000 individuals between 1986 and 2006. This cancer primarily affects those over 65 years, particularly Caucasians and immunocompromised individuals, such as those with chronic lymphocytic leukemia, HIV or organ transplants[3,4].

Gender differences are unclear, though some studies suggest a higher prevalence among women (2:1 ratio). About 35-47% of cases occur in sun-exposed areas like the head and neck, especially the cheeks and eyelids, with 5-10% affecting the eyelids. The remainder are found on the limbs (40%) and torso or genitals (10%). In rare cases, the carcinoma may appear simultaneously in multiple locations, a condition known as merkeliomatosis.

It has been observed that Merkel cell carcinoma tends to occur more frequently in the upper eyelid compared to the lower eyelid, with a distribution of 71% versus 21%. In our case report, the tumor affected the upper eyelid, consistent with this trend. This predilection for the upper eyelid could be related to greater sun exposure or anatomical factors that make this area more susceptible to tumor development. The mass was nodular and exophytic, leading to mechanical ptosis, further emphasizing the tumor’s significant impact on the upper eyelid.

It has been observed[5] that sequences within the genome of Merkel cell carcinoma cells are compatible with a newly identified polyomavirus. This virus may infect carcinoma cells, integrating its genetic material and potentially leading to long-term cell cycle disruption. Although the exact mechanism remains unclear, research has shown that approximately 85% of Merkel cell carcinoma cases contain cells infected with this polyomavirus, strongly supporting the hypothesis that the virus plays a significant role in the development of the carcinoma.

The macroscopic differential diagnosis for Merkel cell carcinoma includes conditions such as chalazia, basal cell carcinoma, nodular angiosarcoma and metastatic tumors. These conditions can mimic the appearance of Merkel cell carcinoma, making it essential to consider a broad range of possibilities during diagnosis to avoid delays in appropriate treatment.

The National Comprehensive Cancer Network (NCCN) in the United States has established guidelines for the diagnostic evaluation of Merkel cell carcinoma (MCC), following a structured approach based on clinical presentation, initial work-up, diagnosis and further evaluation **(Figure 3)**[6,7]. Once MCC is confirmed through clinical and histopathological findings, appropriate staging should be conducted. For patients with palpable lymph nodes, fine-needle aspiration or core biopsy is recommended. In cases with clinically negative lymph nodes, sentinel lymph node biopsy (SLNB) is considered the most accurate method for assessing regional lymph node involvement. SLNB is advised as approximately 25% to 30% of patients with clinically negative nodes are found to have pathological nodal involvement[8].

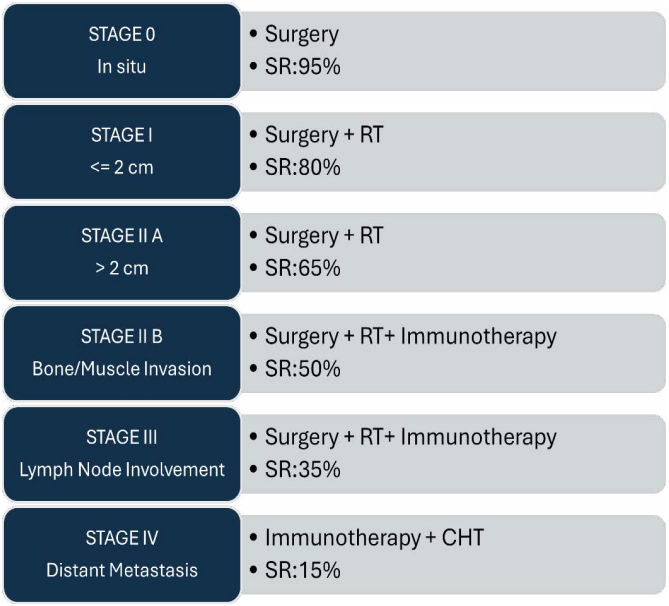

Figure 3: Management of Merkel Cell Carcinoma according to Stage. SR: 5 years survival rate.

Conclusion

In conclusion, early detection and extensive tumor excision with intraoperative margin evaluation, whether through conventional surgery or Mohs micrographic surgery, paired with appropriate eyelid reconstruction, are key to improving survival outcomes in elderly patients with Merkel cell carcinoma of the eyelid. Current evidence supports incorporating sentinel lymph node biopsy during the initial procedure, along with adjuvant radiotherapy. This combined approach not only reduces the risk of local recurrence but also enhances survival by addressing both the primary tumor and any early metastatic spread.

Conflicts of Interest

The authors declare no conflict of interest.

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