Society of Medical Education & Research

2024, Volume 4, Issue 2, Page No: 42-44
ISSN: 3108-4834

Archive of International Journal of Cancer and Allied Science

Merkel Cell Carcinoma: A Case Study and Literature Review

Meryem El Ouazzani[1], Asmaa Lahouaoui[1], Imane Boujguenna[1,2*], Nadia Mansouri[3], Anass Fakhri[1], Hanane Rais[1]

[1]Department of Anatomic Pathology, Med VI University Hospital, Marrakesh, Morocco.
[2]Faculty of Medicine and Pharmacy of Guelmim, Ibn Zohr University, Guelmim, Morocco.
[3]Department of Maxilla-Facial Surgery, Med VI University Hospital, Marrakesh, Morocco.

***E-mail** ✉ imane.boujguenna.fmpg@gmail.com

Abstract

Merkel cell carcinoma (MCC) is an aggressive form of skin cancer that usually occurs in older individuals, often with a history of prolonged sun exposure. This rare malignancy has been associated with various factors, including ultraviolet light, compromised immune function, and the presence of the Merkel cell polyomavirus (MCPyV). Despite ongoing discussions regarding the origin of the cancer cells, this tumor exhibits characteristics similar to the Merkel cells normally found in the skin, both in terms of appearance and immunohistochemical markers. This report describes a case from the Mohamed VI University Hospital Center in Marrakech, where a 79-year-old patient presented with a rapidly growing mass on their upper lip. The diagnosis of MCC relies on comprehensive pathological analysis, including immunohistochemical profiling, and it is essential to differentiate it from other malignancies to ensure proper treatment.

Keywords: Upper lip, Carcinoma, Merkel cell, Pathology

Introduction

Merkel cell carcinoma (MCC) is a rare and highly aggressive malignancy of the skin, often seen in elderly individuals with prolonged sun exposure [1]. Immunosuppression, Ultraviolet radiation, and the Merkel cell polyomavirus (MCPyV) infection are considered significant risk factors contributing to the development of MCC. Despite the ongoing uncertainty about its cellular origin, MCC shares key immunohistochemical markers and structural features with Merkel cells, which are normally present in the skin. This report details a case from the Mohamed VI University Hospital Center in Marrakech, where a 79-year-old patient presented with a rapidly growing mass on their upper lip.

Access this article online https://smerpub.com/

Received: 04 September 2024; **Accepted:** 07 December 2024

How to cite this article: El Ouazzani M, Lahouaoui A, Boujguenna I, Mansouri N, Fakhri A, Rais H. Merkel Cell Carcinoma: A Case Study and Literature Review. Arch Int J Cancer Allied Sci. 2024;4(2):42-4. https://doi.org/10.51847/KesKDsJ7wG

Case study

A 79-year-old male patient, with no remarkable medical history, was observed at the Mohamed VI University Hospital Center in Marrakech, where he presented with a fast-growing mass located on his upper lip. A biopsy was taken, and microscopic analysis revealed an invasive tumor with a variety of growth patterns, including solid sheets, trabecular arrangements, and nests. The tumor cells exhibited moderate size, vesicular nuclei with a distinctive “salt and pepper” chromatin texture, and numerous mitotic figures, along with mitonecrosis. The cytoplasm was scarce and basophilic, and the surrounding stroma was intensely vascularized, showing evidence of lymphovascular invasion. Immunohistochemical tests revealed dot-like staining around the perinuclear region with anti-CK20, positivity for chromogranin and neurofilament, and negative results for anti-CD45 and anti-PS100 antibodies. The tumor grew rapidly, leading to a surgical procedure that

involved wide excision of the tumor and dissection of the nearby lymph nodes **(Figure 1)**.

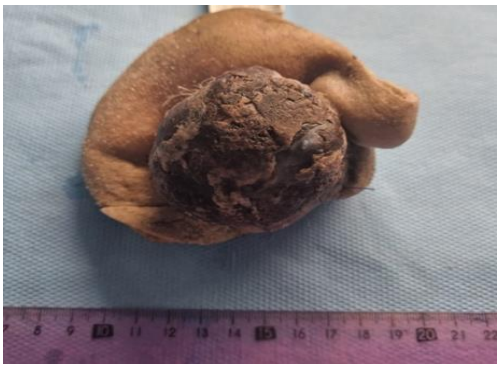

Figure 1. Grossig specimen showing the mass on the upper lip

Results and Discussion

Merkel cell carcinoma (MCC) is recognized for its characteristic appearance of tightly packed blue cells, classifying it as a neuroendocrine tumor. It typically arises within the dermis, often extending into the epidermis, and in some cases, it may invade the deeper subcutaneous tissue. MCC can present with various architectural patterns, such as solid sheets, cellular clusters, or, on occasion, ribbon-like arrangements [2, 3]. The individual tumor cells are often arranged in a way that their borders merge, sometimes resembling lymphocytes, with noticeable mitotic activity and evidence of necrosis. Lymphovascular involvement is frequently observed. Additionally, MCC may show rare features, including squamous or sarcomatoid differentiation, and is commonly associated with adjacent skin lesions like actinic keratosis and in situ squamous cell carcinoma [4].

For an accurate diagnosis and better understanding of the tumor's prognosis, immunohistochemical analysis is essential [5, 6]. A hallmark of MCC is the perinuclear staining pattern with CK20, along with positive immunoreactivity for CD56, chromogranin, synaptophysin, and neurofilament **(Figure 2)** [7].

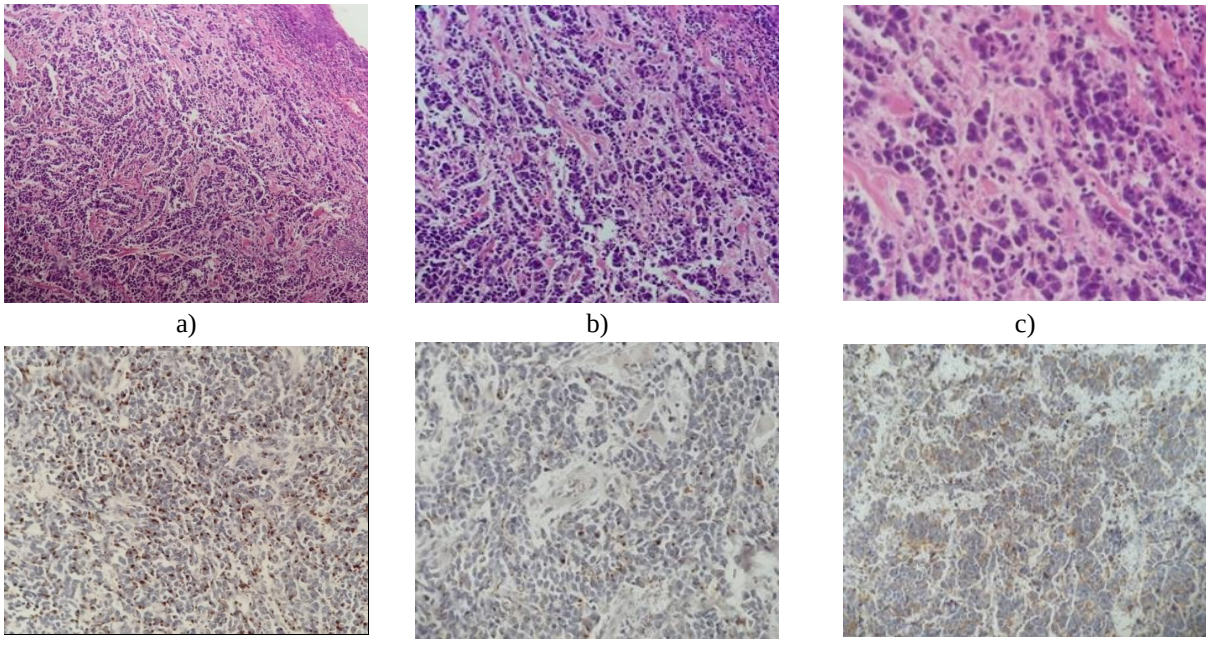

Figure 2. a) the tumor's growth is characterized by formations of nests, trabecular arrangements, and diffuse sheets, b) the cells of the tumor are moderately sized, displaying vesicular nuclei with a "salt and pepper" chromatin pattern, c) staining shows perinuclear dot-like patterns with anti-CK20, d) neurofilament (NF) staining also demonstrates positivity, and e) the tumor cytoplasm shows staining for chromogranin.

When diagnosing Merkel cell carcinoma (MCC), it is important to consider other possible conditions. One such condition is small cell lung carcinoma which can exhibit similar morphological features to MCC. However, the

key difference is that small cell lung carcinoma shows positivity for TTF-1 and lacks CK20 staining, unlike MCC [8]. Another potential misdiagnosis is basal cell carcinoma, as MCC sometimes shares the characteristic of closely packed blue cells. A careful inspection of cellular characteristics is essential, and immunohistochemical testing can provide crucial information for distinguishing the two. Moreover, distinguishing Merkel cell carcinoma from melanoma and squamous cell carcinoma requires a detailed analysis, focusing on both tissue structure and immunohistochemical markers, to ensure an accurate diagnosis [9, 10].

Conclusion

Merkel cell carcinoma is an aggressive form of skin cancer, associated with factors such as UV radiation, immunosuppression, and infection with MCPyV. Accurate diagnosis relies on thorough histopathological examination and immunohistochemical testing. Considering other potential diagnoses is vital for ensuring proper treatment and management.

Acknowledgments: We appreciate the contributions of everyone involved in the care of this patient.

Conflict of Interest: None

Financial Support: None

Ethics Statement: None

References

1. Elder DE, Massi D, Scolyer RA, Willemze R. WHO Classification of Tumours, 4th Ed. Volume 11. 2018. ISBN: 13.978-92-832-2440-2.
2. Bichakjian CK, Lowe L, Lao CD, Sandler HM, Bradford CR, Johnson TM, et al. Merkel cell carcinoma: critical review with guidelines for multidisciplinary management. Cancer. 2007;110(1):1-2.
3. Fraitag S, Moulonguet I. Pathologie cutanée tumorale. Wechsler J, ed. Sauramps médical; 2016. ISBN: 9791030300581
4. Jaeger T, Ring J, Andres C. Histological, immunohistological, and clinical features of Merkel cell carcinoma in correlation to Merkel cell polyomavirus status. J Skin Cancer. 2012;2012:983421.
5. Sheldon JD, Lott Limbach AA. Merkel cell carcinoma of the maxillary sinus: an unusual presentation of a common tumor. Head Neck Pathol. 2021;15(2):691-7. doi:10.1007/s12105-020-01219-y
6. Stanoszek LM, Chan MP, Palanisamy N, Carskadon S, Siddiqui J, Patel RM, et al. Neurofilament is superior to cytokeratin 20 in supporting cutaneous origin for neuroendocrine carcinoma. Histopathology. 2019;74(3):504-13. doi:10.1111/his.13758
7. Lombardi D, Accorona R, Ungari M, Melocchi L, Bell D, Nicolai P. Primary Merkel cell carcinoma of the submandibular gland: when CK20 status complicates the diagnosis. Head Neck Pathol. 2015;9(2):309-14. doi:10.1007/s12105-014-0573-1
8. Pulitzer MP, Brannon AR, Berger MF, Louis P, Scott SN, Jungbluth AA, et al. Cutaneous squamous and neuroendocrine carcinoma: genetically and immunohistochemically different from Merkel cell carcinoma. Mod Pathol. 2015;28(8):1023-32. doi:10.1038/modpathol.2015.60
9. Kervarrec T, Tallet A, Miquelestorena-Standley E, Houben R, Schrama D, Gambichler T, et al. Morphologic and immunophenotypical features distinguishing Merkel cell polyomavirus-positive and negative Merkel cell carcinoma. Mod Pathol. 2019;32(11):1605-16. doi:10.1038/s41379-019-0288-7
10. Gauci ML, Aristei C, Becker JC, Blom A, Bataille V, Dreno B, et al. Diagnosis and treatment of Merkel cell carcinoma: European consensus-based interdisciplinary guideline–Update 2022. Eur J Cancer. 2022;171:203-31. doi:10.1016/j.ejca.2022.03.043