

## سرطانة خلية ميركل في الجلد\_ تقرير حالة مع مراجعة الأدبيات

د. فريز أحمد<sup>1</sup> ، د. صفاء قطليش<sup>2</sup>

<sup>1</sup> مدرس في قسم الباثولوجيا والباثولوجيا الخلوية - كلية الطب البشري - جامعة دمشق.

<sup>2</sup> Ph.D في الباثولوجيا والباثولوجيا الخلوية - كلية الطب البشري - جامعة دمشق.

Email: [safaa.qatleesh@damascusuniversity.edu.sy](mailto:safaa.qatleesh@damascusuniversity.edu.sy)

### الملخص:

خلفية البحث وهدفه: خباثة نادرة شديدة العدوانية ضمن الجلد مع إنذار سيء. تحدث بشكل وصفي في الجلد المعرض لأشعة الشمس.

مواد البحث وطرائقه: تقرير حالة لدى ذكر بالغ من العمر 104 سنة مع عقيدة مؤلمة تقليدية جلدية على الأنف تضخمت بشكل سريع. أجري لها استئصال والموجودات الباثولوجية أكدت التشخيص كونها سرطانة خلية ميركل باستخدام الكيمياء النسيجية المناعية ونفت جميع الآفات الأخرى.

النتائج: هذه الحالة والمراجعة الأدبية لها شرحت الجوانب السريرية والباثولوجية لمرضى سرطانة خلية ميركل وهذا قد يساهم بالتعرف على المرض بشكل باكر وعلاج هذه الأورام.

كلمات مفتاحية: سرطانة خلية ميركل، خلايا غدية ضماوية عصبية، الجلد، CK20

تاريخ الايداع: 2022/4/7

تاريخ القبول: 2022/5/31



حقوق النشر: جامعة دمشق -

سورية، يحتفظ المؤلفون بحقوق

النشر بموجب CC BY-NC-SA

## Merkel Cell Carcinoma of Skin-Case Report with Literature Review

Dr.Fariz Ahmad<sup>1</sup>, Dr.Safaa Qatleesh<sup>2</sup>

<sup>1</sup> Professor in the Department of Pathology- Faculty of Medicine - Damascus University.

<sup>2</sup> Ph.D in Pathology - Faculty of Medicine- Damascus University.

E-mail: [safaa.qatleesh@damascusuniversity.edu.sy](mailto:safaa.qatleesh@damascusuniversity.edu.sy)

Received: 7/4/2022

Accepted: 31/5/2022



**Copyright:** Damascus University- Syria, The authors retain the copyright under a CC BY- NC-SA

### Abstract

**Background & Aim:** Merkel cell carcinoma is a rare, very aggressive malignancy of the skin with a poor prognosis. It is typically located on sun-exposed skin.

**Materials & Methods** Herein, we report a 104 - year - old male with a painful, ordinary

skin lesion on nose that enlarged rapidly. Total excision was done. Pathologic findings confirmed diagnosis of Merkel cell carcinoma by immunohistochemistry analysis and rule out other lesions.

**Results:** This case report and literature review demonstrate the clinical and histopathologic aspects of

MCC, which will help in recognizing and treating these tumors early.

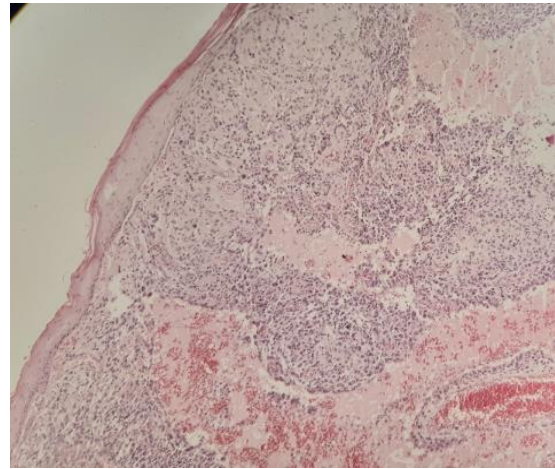
**Key Words:** Merkel Cell Carcinoma, Neuroendocrine Cell , Skin, CK20.

## 1. Introduction

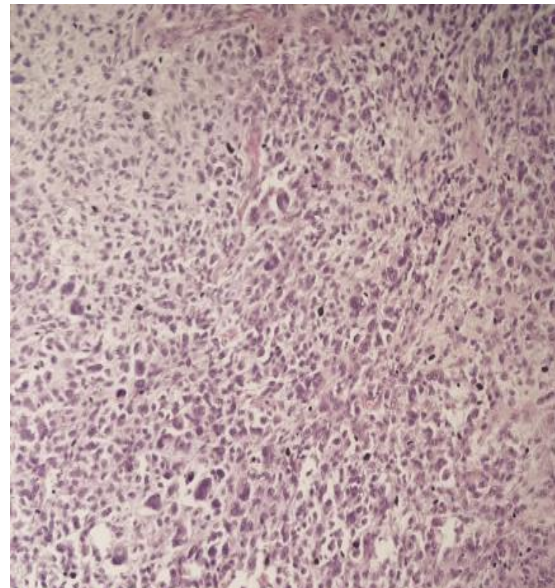
Merkel cell carcinoma (MCC) is a rare, malignant neuroendocrine tumor of skin (Cornejo & Miller, 2019). It usually presents as a ordinary-appearing lesion occurs mainly in bodily areas exposed to sunlight, such as the face, neck, and limbs, with low incidence and high recurrence. The diagnosis is rarely suspected at the time of biopsy and the clinical differential diagnosis includes common entities, such as basal cell carcinoma (BCC), epidermoid cyst, pilar cyst, metastasis of squamous cell carcinoma or even amelanotic melanoma (Heath *et al.*, 2008). MCC often presents as a rapidly growing, asymptomatic, bluish -red dermal nodule or papule that develops during weeks to months (Heath *et al.*, 2008). MCC characterized by local recurrence, early involvement of the regional lymph nodes and rapid distant metastases (Metzinger, Wolfer, Disa, Kupersmith, & Robertson, 2000). Due to its aggressiveness and benign clinical appearance, the prognosis of this neoplasm is poor (Arnold, Odom, Andrews, & James, 1990; Metzinger *et al.*, 2000). Immunohistochemical staining plays an important role in the early diagnosis (Bielamowicz, Smith, & Abemayor, 1994; Cornejo & Miller, 2019).

## 2. Case Report

This case concerns an 104-year-old man who presented with a 3.5×3.0×2.2 cm red nodule on nose, which was initially diagnosed as basal cell carcinoma at the beginning of January 2020 with no significant past medical history presented to the surgery clinic. Surgical resection was performed at Mouwasat University Hospital. The results of pathological examination after surgery showed small cell malignant tumor of about 3.5×3.0×2.2 cm (Figure 1) Histologically, the tumor composed of round cells with scanty cytoplasm formed in diffuse pattern (cohesive cells) with vesicular nuclei with abundant mitotic figures, rare bizarre cells noted (Figure 2), invading the overlying skin with broad ulceration.



**Figure 1: Small round blue cells infiltrate in the dermis invading the lamina (Low magnification).**



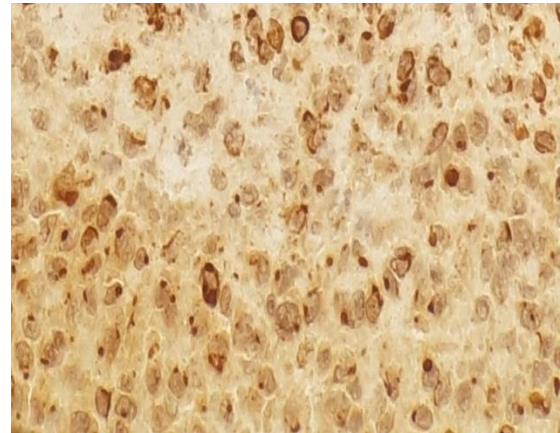
**Figure 2: Small round blue cells infiltrate in the dermis invading the lamina (High magnification).**

The immunohistochemistry analysis in our hospital showed that the tumor was negative for CK7, CD20, CD3, HMB45, and CK5/6 but positive for CK20, consisting with MCC.

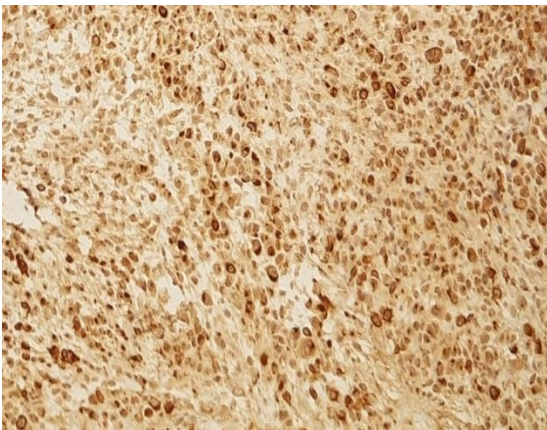
The CK20 positive immunohistochemical staining demonstrating paranuclear dot-like staining (IHC × 20) (Figure 3,4,5,6) was diagnostic and an indicator for Merkel cell carcinoma. However, the other negative staining ruled out other lesions, which gives the same morphology by routine H&E stain.



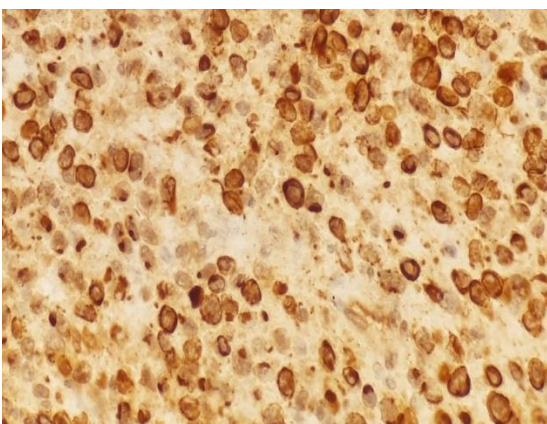
**Figure 3: Antibody CK20 staining by immunohistochemistry analysis (low magnification with internal control).**



**Figure 6: Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis (High magnification).**



**Figure 4: Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis (Low magnification).**



**Figure 5: Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis.**

According to the pathology and immunohistochemistry, this patient was diagnosed with MCC.

Treatment consisted of surgical excision of the tumor with a wide margin as well as chemotherapy.

### 3. Discussion

MCC originates in the dermis, but can expand into the epidermis (Uchi, 2018). The tumor is composed of sheets of small round blue cells with marked nuclei. The cells have frequent mitoses with variegated “salt and pepper” chromatin (Wang, Byrne, Jacobs, & Taube, 2011). Immunohistochemistry (IHC) is used for the final diagnosis of MCC. CK20 stains in a paranuclear dot-like pattern in 80–90% of MCC cases due to clumping of intermediate filaments in the cytoplasm. MCCs frequently stains positive for neuroendocrine markers, such as chromogranin, synaptophysin and CD56. MCC stains negative for TTF-1, CD45, and HMB45 which distinguishes it from small cell lung carcinoma lymphoma and melanoma, respectively (Schmerling et al., 2018; Uchi, 2018; Wang *et al.*, 2011).

### 4. Conclusion

This case report and literature review demonstrated the clinical, behavioral and histopathologic aspects as well as features of MCC, which will help in recognizing and treating these tumors earlier.

**Table(1): for all figures:**

Figure(1)	<b>Small round blue cell infiltrate in the dermis invading the lamina (Low magnification).</b>
Figure(2)	<b>Small round blue cells infiltrate in the dermis invading the lamina (High magnification).</b>
Figure(3)	<b>Antibody CK20 staining by immunohistochemistry analysis (low magnification with internal control).</b>
Figure(4)	<b>Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis (Low magnification).</b>
Figure(5)	<b>Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis.</b>
Figure(6)	<b>Antibody CK20 paranuclear dot-like pattern staining by immunohistochemistry analysis (High magnification).</b>

**References:**

1. Arnold, H. L., Odom, R. B., Andrews, G. C., & James, W. D. (1990). *Andrews' diseases of the skin: clinical dermatology*: WB Saunders Company.
2. Bielanowicz, S., Smith, D., & Abemayor, E. J. T. L. (1994). Merkel cell carcinoma: an aggressive skin neoplasm. 104(5), 528-532.
3. Cornejo, C., & Miller, C. J. J. D. C. (2019). Merkel cell carcinoma: updates on staging and management. 37(3), 269-277.
4. Heath, M., Jaimes, N., Lemos, B., Mostaghimi, A., Wang, L. C., Peñas, P. F., & Nghiem, P. J. J. o. t. A. A. o. D. (2008). Clinical characteristics of Merkel cell carcinoma at diagnosis in 195 patients: the AEIOU features. 58(3), 375-381.
5. Metzinger, S. E., Wolfer, R. S., Disa, J. J., Kupersmith, J. E., & Robertson, B. C. J. S. m. j. (2000). Recurrent Merkel cell carcinoma of the upper extremity. 93(3), 340-345.
6. Schmerling, R. A., Casas, J. G., Cinat, G., Ospina, F. E. G., Kassuga, L. E., Tlahuel, J. L. M., & Mazzuocolo, L. D. J. J. o. G. O. (2018). Burden of disease, early diagnosis, and treatment of Merkel cell carcinoma in Latin America. 4.
7. Uchi, H. J. F. i. o. (2018). Merkel cell carcinoma: an update and immunotherapy. 8, 48.
8. Wang, T. S., Byrne, P. J., Jacobs, L. K., & Taube, J. M. (2011). Merkel cell carcinoma: update and review. Paper presented at the Seminars in cutaneous medicine and surgery.