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| RESEARCH ARTICLE

Merkel Cell Carcinoma of Atypical Location: Case Report and Review of the Literature

MOUJAHID Asmaa[1] ✉ SAGNON Indou[2], BADAOUI Zakaria[3], RIBAG Yassamina[4], KHALES Amine[5], ACHBOUK Abdelhafid[6] and EL KHATIB Karim[7]

[1234567]Department of Plastic, Reconstructive and Burn Surgery, Mohamed V Military Training Hospital, Faculty of Medicine and Pharmacy, Mohamed V University, Rabat - Morocco

Corresponding Author: MOUJAHID Asmaa, **E-mail**: asmaamouj@gmail.com

| ABSTRACT

Merkel cell carcinomas are rare and aggressive skin tumours, with a high risk of local, lymph node and metastatic recurrence, and a guarded prognosis. Their preferred location is in sun-exposed areas. Surgery remains the cornerstone of treatment, with recommended lateral margins of up to 3 cm, including the underlying fascia, in order to limit the risk of recurrence. Adjuvant radiotherapy improves local and regional survival, given the radiosensitivity of these carcinomas. We report an atypical case of Merkel cell carcinoma in the gluteal region, managed by a multidisciplinary team combining surgery and radiotherapy.

| KEYWORDS

Merkel cell carcinomas, Localization, Surgery, Radiotherapy.

| ARTICLE INFORMATION

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1. Introduction

Merkel cell carcinoma was first described by Toker as "trabecular carcinoma of the skin". It is a rare and highly aggressive primary skin tumour, occurring preferentially in areas exposed to light [1-4].

Merkel's carcinoma mainly affects the elderly. Two main aetiologies are incriminated in its occurrence: integration of Merkel polyomavirus (MCPyV) and/or chronic UV exposure. A tumour with a high rate of local or lymph node recurrence, it may present distant metastases and has a high mortality rate, with a median survival of 9 months. Chemotherapy used to be the first-line treatment for metastatic Merkel cell carcinomas, but new recommendations have emerged to improve the management of these carcinomas [5-9].

The interest of our work lies essentially in the atypical location of Merkel cell carcinoma.

2. Clinical case

The patient was 62 years old, with no notable pathological history, and had been presenting for three months with a subcutaneous swelling in the left gluteal region, firm and slightly tender, with no ulceration opposite, measuring 2 cm long. A first biopsy-exeresis was performed, without margin, concluding to a morphological and immunohistochemical aspect of a Merkel cell carcinoma.

Given the atypical location of the carcinoma, the patient was presented at a multidisciplinary consultation meeting, during which an extension assessment was requested, as well as a repeat operation.

The patient underwent lymph node ultrasound of the abdominal, pelvic and inguinal regions and a bone scan, which came back without any particularities, resulting in this carcinoma being classified as stage I.

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During surgery, we performed a revision with 2cm margins laterally and with deep fascia, and additional reconstruction during a second operation, after histological results confirming total tumour removal. We opted for a rotation flap to reconstruct the resulting loss of tissue. The post-operative course was straightforward, with no complications (Fig. 1-3).

Two weeks after removal of the sutures, the patient underwent adjuvant radiotherapy sessions for additional management.

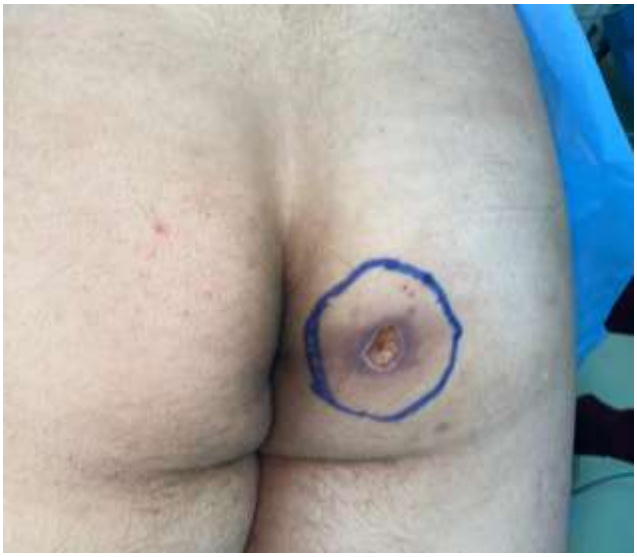

Figure 1: Clinical aspect of post-biopsy loss of substance following excision of Merkel cell carcinoma - Revision with 2cm margins

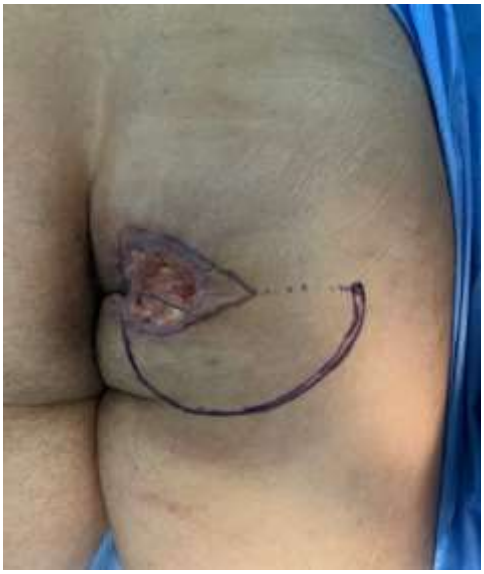

Figure 2: Clinical aspect of coverage of loss of substance by a rotation flap

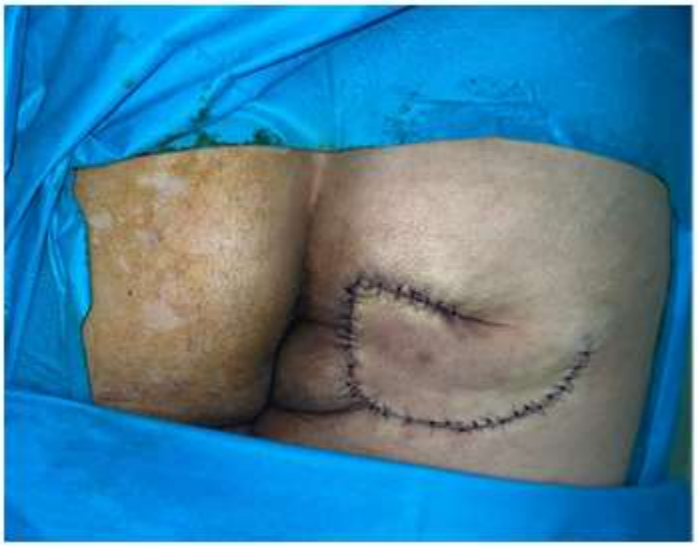

Figure 3: Immediate intraoperative clinical aspect

3. Discussion

Merkel cell carcinomas are rare and aggressive primary cutaneous neuroendocrine tumours, and their incidence is increasing worldwide, with an estimated 3250 cases per year in 2025 [10-14].

Numerous studies have described the two main etiologies involved in the pathophysiology of Merkel cell carcinoma: Merkel polyomavirus (MCPyV) and/or chronic UV exposure [15-20].

Exposure to the sun is the main reason for this, given the frequent location of these carcinomas in exposed areas. Merkel's carcinomas are also considered to be synchronous or metachronous tumours, whether skin tumours or malignant tumours involving other organs [21-25].

The management of Merkel cell carcinoma requires a multidisciplinary consultation, particularly as there is no clear consensus on the best approach, given the rarity of these tumours and the virtual absence of randomised prospective studies. Surgery remains the cornerstone of treatment, with a discussion concerning the margins of exeresis, which vary between 2 and 3cm laterally depending on the location of the carcinoma and its size, and in depth by taking away the underlying fascia. Coverage is imminent, to allow complementary management by radiotherapy. Lymph node dissection remains a subject of debate; some authors recommend dissection only in the presence of poor prognostic factors; other studies recommend sentinel lymph node biopsy to determine whether or not lymph node dissection is indicated. Merkel cell carcinoma is radiosensitive, and several studies have shown the efficacy of radiotherapy at the tumour site, in terms of survival rates and regional and distant recurrence. Chemotherapy, on the other hand, is only indicated in cases of locally advanced Merkel cell carcinoma or metastatic tumour [21,26-32].

In our patient, the Merkel tumour was located in the left gluteal region. The preferred location for Merkel cell carcinomas is open areas exposed to the sun [26,31-33]. Following a multidisciplinary consultation meeting, the treatment decision was surgical resection with adjuvant radiotherapy and coverage, which improved the patient's survival. Inguinal lymph node dissection was not performed, and sentinel lymph node testing was not performed, as there were no signs of a poor prognosis. In all cases, clinical and radiological monitoring was instituted.

4. Conclusion

Merkel cell carcinomas are rare and aggressive tumours, occurring mainly in the extremities and exposed areas. In our work, we present the case of a Merkel cell tumour of atypical location, in the gluteal region. Treatment is still optimal: after multidisciplinary consultation, surgery is the cornerstone of treatment, combined with adjuvant radiotherapy. However, new therapies have emerged in recent studies, such as immunotherapy for aggressive metastatic forms, or targeted and cellular therapies [9].

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