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Case Report

Merkel Cell Carcinoma as a Diagnosis of an Inguinal Mass, a Case Report

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Abstract

Introduction: Merkel cell carcinoma is a rare and aggressive cutaneous Neuroendocrine tumor. Overall survival is related to different factors. The mainstay of treatment is surgery and in some cases it may be accompanied by radiotherapy. Chemotherapy is limited to metastatic disease.

Case Report: A 64-year-old man presented with a mass in the left inguinal region with a biopsy compatible with Merkel Cell Carcinoma. The primary lesion was not found and no systemic dissemination. The patient was treated with local surgery and adjuvant radiotherapy. No signs of recurrence seen after 6 months of follow up.

Discussion: Unknown primary MCC is described in the literature in a small number of cases. The most common site of presentation is the inguinal lymph node area. Tumors of unknown primary sites are associated with a decreased risk for distant metastasis and overall mortality; also they have significantly low incidence of recurrence. They have better overall survival compared with patients with known MCC and affected lymph nodes. Chemotherapy has not demonstrated improved outcomes in these patients.

Conclusions: Although MCC is a rare tumor, especially with unknown primary lesion. The gold standard of treatment is a multidisciplinary management. Unknown primary MCC has better prognosis compared with MCC with known primary lesion.

Keywords: *Merkel Cell Carcinoma; Unknown Primary Lesion; Lymph Node Affected*

Abbreviations

MCC: Merkel Cell Carcinoma; CT: Computerized Tomography; PET: Positron Emission Tomography

Introduction

Merkel cell carcinoma is a rare and aggressive cutaneous Neuroendocrine tumor affecting mainly head and neck [1,2]. It is the second most common cause of skin cancer death after melanoma [3,4].

The diagnosis is in the elderly (65 - 75 years) and the main risk factors are Caucasian, males, chronic ultraviolet light exposure and immunosuppression [4]. Recently, polyomavirus has been identified as a risk factor, although its mechanism of action is unclear [5].

The diagnosis has to include a thorough examination of the skin and regional lymph nodes and an imaging test as a computerized tomography (CT) or a positron emission tomography (PET) with 18-fluorodeoxyglucose [2]. Classification stage is summarized in table 1.

Stage		Number of cases	5-year survival
Stage I	Primary tumor size, ≤ 2 cm	Local disease 60 - 70%	60 - 80%
Stage II	Primary tumor size, > 2 cm		
Stage III	Nodal disease	30%	50%
Stage IV	Metastases beyond the regional lymph basin	5 - 10%	20%
Overall			40%

Table 1: *Merkel cell carcinoma staging, number of cases and 5-year survival.*

Overall survival is related to different factors, the unfavorable ones are age > 70, male sex, trunk site, head and neck site (especially lip), size of primary > 2 cm, initial nodal disease presentation (especially if > 2 involved nodes), initial distant disease presentation and some points of the histology (small-cell size, high mitotic rate, depth of invasion) lymphovascular invasion, infiltrative rather than a nodular growth pattern [1,2,6].

The mainstay of treatment is surgery and in some cases it may be accompanied by radiotherapy. The main indications for adjuvant radiotherapy are primary tumor > 2 cm, positive resection margin or margins in risk, lymphovascular invasion, extracapsular extension of tumor and documented regional lymph node involvement. Chemotherapy is indicated in metastatic disease there is no clear evidence about its role to prevent recurrence [2,7].

Case Report

A 64-year-old man was referred to our center in Spain, for a mass in the left inguinal region of two months duration. An ultrasound has been done, identifying a 4 x 4 cm solid hypoechogenic mass and another lesion of 12 x 10 mm, that were suspicious of pathologic lymph nodes. No other lesion found in the physical exploration and the first probable diagnosis was a lymphoma, so a biopsy of the bigger mass was made by tru-cut. The biopsy was compatible with Neuroendocrine carcinoma with extended tumoral necrosis and the immunohistochemistry (positive for CKAE1-AE3, neuron-specific enolase, chromogranin, synaptophysin, CK-20 and negative for CD3, CD20, S100, CK7, TTF-1, CDX-2 Gata3, PSA and SOX10) compatible with Merkel Cell Carcinoma (MCC).

The patient was explored exhaustively and no dermal lesions were identified. To complete the study a PET-CT was performed, identifying only the two lesions found previously in the ultrasound, no other findings. The lesions are shown in figure 1 and 2.

The patient had a lymph node metastasis of MCC located in the inguinal region without primary lesion, so it was a stage IIIa (8th Edition American Joint Committee on Cancer Merkel cell staging system) [1]. It was commented in the committee and the resection of the lesion and adjuvant radiotherapy was decided. The surgical technique was a left inguinal lymphadenectomy. The two known lesions were found and the rest of the lymph nodes were negative (2/12 lymph nodes). After that, the patient received radiotherapy (66Gy). No other treatments. No signs of recurrence 6 months after the surgery.

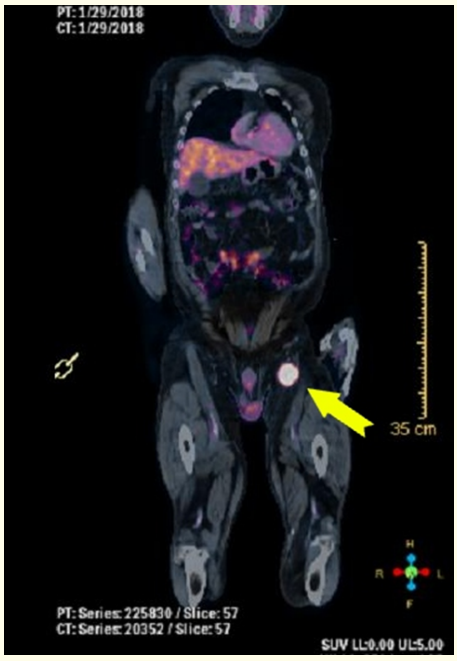

Figure 1: *Sagittal PET image. Arrow indicates the inguinal mass.*

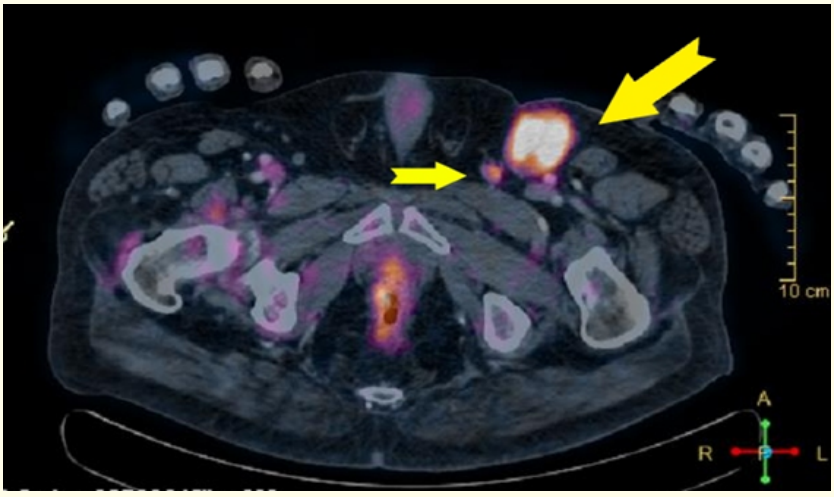

Figure 2: *Axial PET image. Arrows indicate the affected area, the two suspicious nodules (bigger one has a SUV max of 8.1 and the other one has a SUV of 4.2). The uptake at the rectum is normal due to the digestive elimination of the marker.*

Discussion

Unknown primary MCC is described in the literature in a small number of cases and the most common site of presentation is the inguinal lymph node area. It represents 5% of all MCC patients and between 32-40% of those with an affected nodal disease [6,8-11].

Tumors of unknown primary sites were associated with a decreased risk for distant metastasis and all-cause mortality, also they have significantly improved recurrence [6,8]. These patients had better overall survival compared with patients with known MCC and affected lymph nodes [6,8-12]. The explanation is not clear, although it could be associated with improved cell-mediated immunity, which clears the primary tumor and targets residual disease [12]. In the 8th Edition AJCC staging system of MCC, the unknown primary tumors were classified as stage IIIa, instead of IIIb as in the previous edition [1].

This has a special interest because of unknown primary site MCC patients not necessary need aggressive treatments, since, there are studies that not associated the chemotherapy with improved outcomes in these patients [8,14].

Conclusion

Merkel cell carcinoma is a rare and aggressive disease that can appear without primary lesion. It is important, not just a complete exploration, but a A PET or CT are necessary to complete assessment before treatment. The gold standard treatment is a multidisciplinary management where the surgery is the basis of the treatment, and in selected patients the radiotherapy has to be used as adjuvancy. Unknown primary MCC has better prognosis compared with MCC with known primary lesion. In these cases, chemotherapy has not demonstrated improved outcomes.

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