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Case Report

Merkel Cell Carcinoma of the Inguinal Lymph Node in the Absence of a Primary Site: A New Case Report and Literature Review

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Abbreviations MCC: Merkel Cell Carcinoma; LN: Lymph Node

Abstract

Introduction: Merkel Cell Carcinoma (MCC) is a rare and aggressive neuroendocrine tumor of the skin. The main characteristics are frequent local recurrences and disseminations to regional lymph nodes and distant organs. MCC within the lymph nodes in the absence of a primary site is rare and few cases have been reported by the literature.

Case Report: We report a case of MCC presenting as a painless mass in the left inguinal area for 6 months in a 48-year-old women. The histopathology of the excised lesion revealed a poorly differentiated basophilic small cell tumor. The immunohistochemical study finding the diagnosis of a metastatic MCC. Despite extensive clinical and radiological investigation, we failed to identify the origin of the tumor.

Conclusion: Rare cases of MCC confined to a lymph node without an apparent primary site have been reported. We report a new case of MCC in the inguinal lymph node without identification of the primary site.

Introduction

Merkel Cell Carcinoma (MCC) was first described by Toker in 1972 as "trabecular carcinoma of the skin" [1]. This aggressive tumor recurs locally and frequently disseminates to regional lymph nodes and distant organs, primarily to the liver and bones. However it has been previously reported in various anatomical sites [2], Lymph Node (LN) metastatic MCC in the absence of a primary site is extremely rare [3] and for this reason there is no standard approach to its management.

In the present study we report the case of a Moroccan patient with LN metastatic MCC in the absence of a primary site and we described the clinical characteristics, natural history, alternative hypotheses and pertinent therapy of this uncommon tumor.

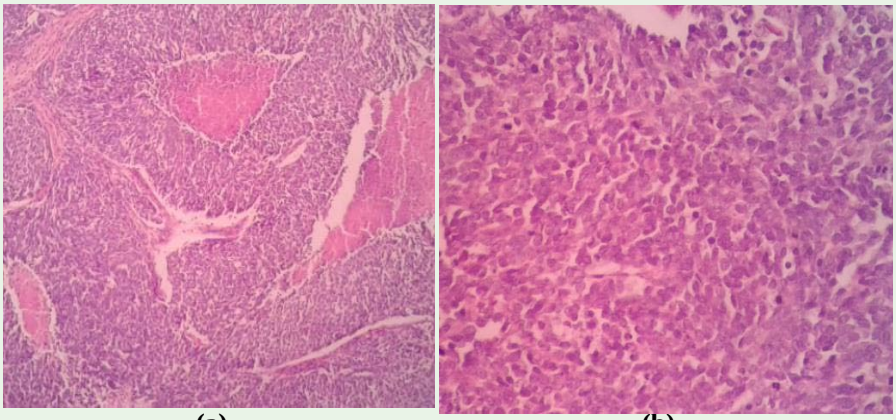
Case report

A 48-year-old woman presented with a painless, 10-cm palpable mass of 6 months duration in the left inguinal area. No suspicious skin lesions were identified and the patient denied any previous treatment. The laboratory values were normal. The ultrasound findings suggested a conglomerated lymph node in the left inguinal area. An excision biopsy was performed.

Histopathologically, the normal lymph node tissue was replaced by diffuse infiltration of undifferentiated tumor cells. They were monomorphic, basophilic, small, round cells with ovoid vesicular nuclei and scanty cytoplasm. Numerous mitotic figures were observed (Figure 1a, 1b). Immunohistochemically, the tumor cells were positive for cytokeratin 20 in a dot-like pattern (Figure 2a) and in a membranous pattern for CD56 along the cell borders (Figure 2b). The tumor cells were negative for cytokeratin 7 and CD45. These findings were consistent with the diagnosis of a metastatic MCC. Computed tomography of the chest and abdomen, was unremarkable. The results of the upper and lower gastrointestinal endoscopy were within normal limits.

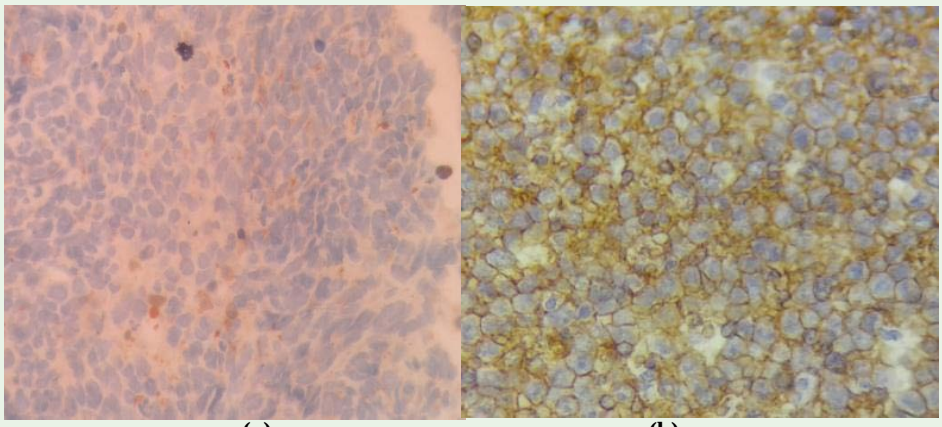
The inguinal mass was excised and sent for pathology department for histological evaluation. At macroscopic examination, the specimen has corresponded a tumoral mass, measuring 10x6 cm, Lobulated and white-greyish, firm consistency with necrotic remodeling (Figure 3).

The histological examination confirmed the diagnosis of a metastatic MCC in inguinal lymph node. The patient is currently being treated with adjuvant radiation therapy and is scheduled to receive systemic chemotherapy.



(a) (b)

Figure 1: Morphologic features of nodal Merkel cell carcinoma.
1(a): Nested and organoid arrangements with Sheets of growth pattern and geographic necrosis (Hematoxylin & eosin, 10x).
1(b): Tumor composed with basophilic, small, round, monomorphic tumor cells with scanty cytoplasm, along with conspicuous mitotic figures (Hematoxylin & eosin, 20x).



(a) (b)

Figure 2: Immunohistochemical staining of nodal Merkel cell carcinoma.
2(a): Tumor cells are focal positive for CK20 with characteristic perinuclear 'dot-like' staining.
2(b): Tumor cells also express synaptophysin.

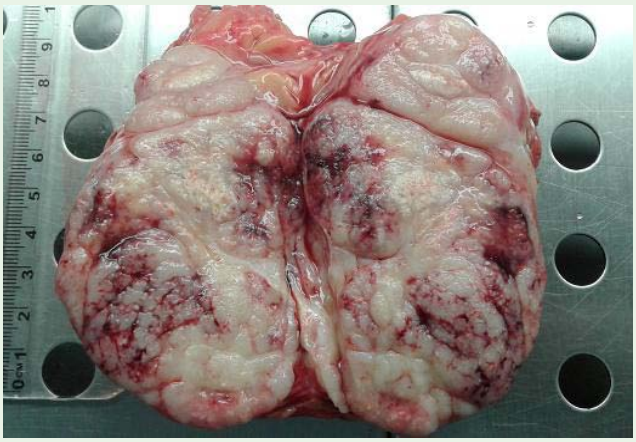


Figure 3: Gross appearance of conglomerated lymph node showing a lobulated and white- greyish mass.

Discussion

Merkel Cell Carcinoma (MCC) is an uncommon and aggressive neuroendocrine tumor occurring most commonly on the head and neck in elderly Caucasian patients. MCC was first described in 1972 [1].

Etiology of MCC is thought to be related to ultraviolet exposure and a recently discovered polyomavirus (Merkel cell polyomavirus) [4].

The most frequent sites of metastasis are distant lymph nodes, distant skin, CNS, and bone. The histological diagnosis can be difficult, because with the conventional light microscopy Merkel cell carcinoma can be misdiagnosed as any other poorly differentiated small cell neoplasm including metastatic melanoma, lymphoma, small cell carcinoma, and neuroendocrine carcinoma metastatic from other organs. Immunohistochemical staining play an important role in the early diagnosis [5-9].

The most common staging system is that described by Yiengpruksawan et al. stage I disease for isolated local lesion, stage II disease is characterized by metastatic spread to regional lymph nodes and stage III has evidence of distant metastatic disease at the time of initial presentation [9].

Merkel cell carcinoma of the lymph node without an obvious primary site has been rarely reported. Tarantola et al. reported 23 cases of MCC with no evidence of a primary site; in this study the most common site of presentation for unknown primary MCC was the inguinal lymph node basin [10].

Two alternative hypotheses for this rare entity have been proposed. First, the lymph node is the primary site, rather than the metastatic one. This hypothesis is generally not accepted because Merkel cells or their precursors have never been identified in lymph nodes, although the possibility of an anomalous acquirement of neuroendocrine characteristics upon malignant transformation of immature totipotent stem cells cannot be ruled out. Second, the presence of these malignant processes in lymph nodes may be the result of metastatic spread from an occult or regressed primary carcinoma [11].

The rarity of this phenomenon makes it very difficult to set standards for treatment. Aggressive surgical treatment remains the mainstay of therapy with adjuvant radiation therapy for local control. Despite the limited effectiveness of chemotherapy, adjuvant chemotherapy can be considered on an individual basis [12,13].

Conclusion

Lymph node metastatic MCC in the absence of a primary site is extremely rare. In the literature, the most common site of presentation for unknown primary MCC was the inguinal lymph node basin. The rarity of this phenomenon makes it very difficult to establish treatment standards.

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