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Case Report

# Very rare localization of a merkel cell carcinoma: about a case and review of the literature

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**Abstract:** We report a case of Merkel cell carcinoma in a 34-year-old woman in the form of a subcutaneous nodule measuring 5 x 3 x2 cm from the right thigh root. Wide surgical excision is the only treatment performed. After a follow-up of 10 months, the patient is still alive without metastasis or recurrence. The review of the literature tells us that the origin of this tumor remains hypothetical and that only the immunohistochemical study provides diagnostic confirmation in particular the coexistence of Neurofilament and cylokeratin. The specific enolase neuron and membrane epithelial antigen are consistently positive. Overall, the prognosis is poor: the survival rate at 3 years is 62%. Certain parameters correlated with the prognosis are specified.
**Keywords:** Merkel; carcinoma; Root of the thigh

## INTRODUCTION

Neuroendocrine cutaneous carcinomas (CNECs) are rare malignant tumors, originally described by Tocker in 1972 [1] as trabecular carcinoma of the skin. The term Merkeloma or "Merkel cell tumor" is proposed in 1975 by Tang and Tocker [2] in the presence of neuro secretory grains in tumor cells very similar morphologically to those observed in normal Merkel cells under electron microscopy. The Merkel cell is normally found in the basal layer of the epidermis.

Despite the unmistakable resemblance between the Merkel cell and the neuroendocrine cutaneous carcinoma cells, there is no morphological or phytogenetic argument to establish the filiation between the normal Merkel cell and neoplasia so that the origin of cutaneous neuroendocrine carcinoma remains Enigma [3]. The case of CNEC that we report, illustrated by the data of the literature, will allow us to put the point on this rare cutaneous tumor

## CASE REPORT:

Mrs.M.N, Aged 35, with no significant antecedents, consults in January 2016 for a subcutaneous nodule in the root of the right thigh appeared 5 months ago which gradually increased in volume. On examination, the nodule is of normal coloring, of firm consistency, and is seated at the level of the antero-internal surface of the right thigh. The ganglionic areas are free (Fig 1). The radiological study by MRI of the right thigh showed that it was a tissue mass of the right inguinal fold extended to the anterior surface of the right thigh evoking a sarcoma of the soft parts (FIG. 2)

Histopathologic analysis of a biopsy fragment concluded with cutaneous neuroendocrine carcinoma, the diagnosis is confirmed by the immunohistochemical study. The patient underwent extensive surgical excision under loco-regional anesthesia. With lymph node dissection of the satellite ganglia (FiG 3). On macroscopic examination, it is a skin and subcutaneous plaque, measuring 2x2, 5x1 cm; the epidermis is not spared (FiG 4). The histopathological analysis of the coin finds the same data of the biopsy and specifies that the limits of lateral and deep excisions are unharmed. No adjuvant therapy is performed. After a 10-month follow-up, the patient is still alive without recurrence or metastasis.

**Iconography**

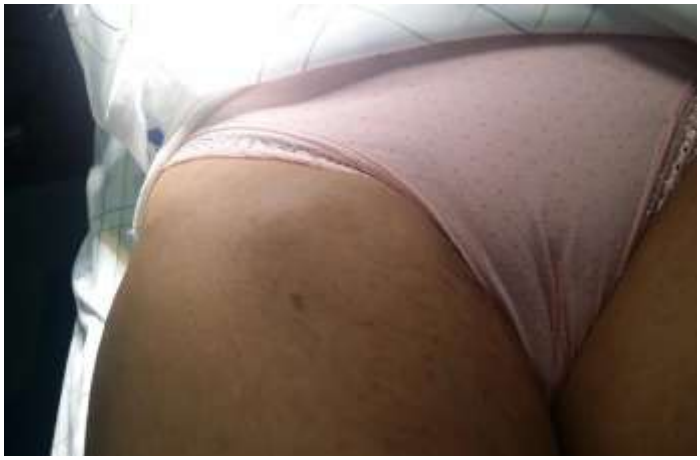

**Fig 1: tumefaction at the root of the thigh Right without skin lesion**

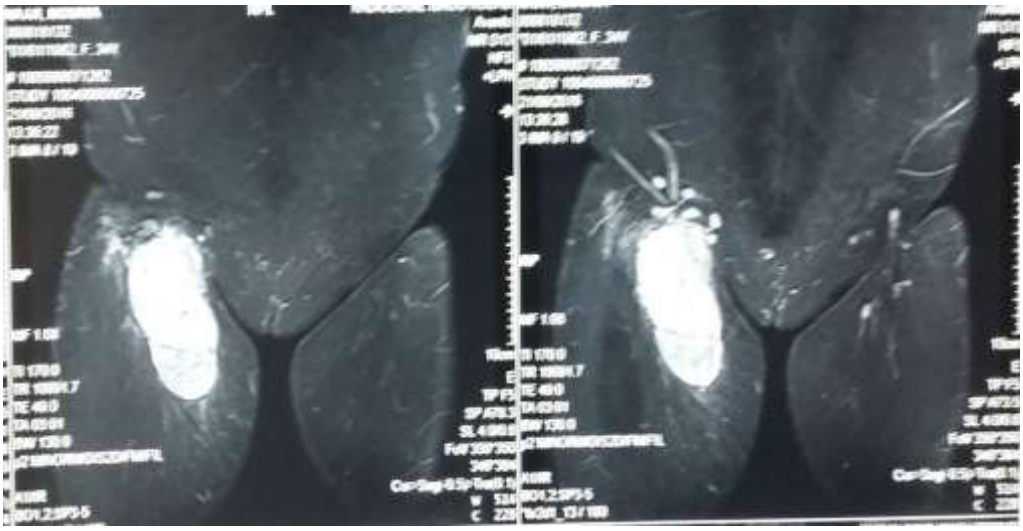

**Fig 2: IRM of the right thigh showing the tumor**

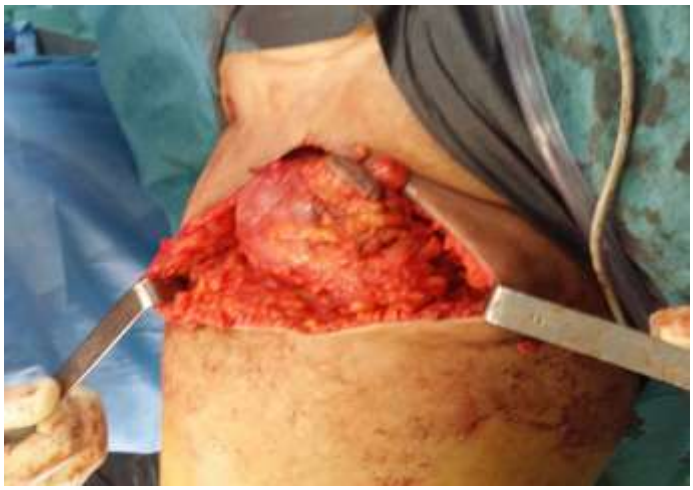

**Fig 3: Peroperative image showing tumor resection**

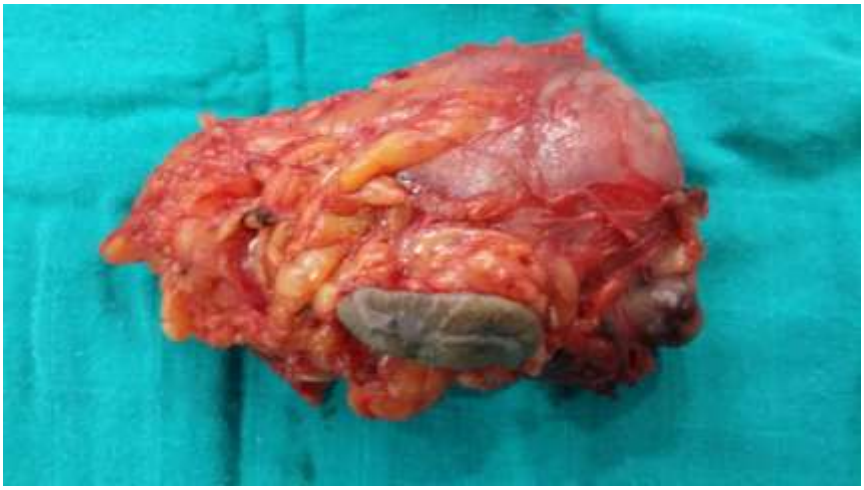

**Fig 4: Image showing the operating room**

**DISCUSSION:**

The reputation of scarcity of the CNEC seems relative since Halioua and Ortonne [4] count 450 cases over a period of 15 years. This tumor affects the female adult, the average age of onset is 60 years (15-92 years) [5], and no case is described in children. The tumor affects almost exclusively white subjects. Clinically, it is in the form of a nodule or a cutaneous or subcutaneous plaque

The average diameter is 2 cm. globally well limited, unencapsulated and mobile with respect to the underlying, painless, non-pruriginous, mostly violet-red [3] and sparingly epidermal planes. Some sites are preferentially interested: the face (more than 50%), the thorax and very rarely the members it generally respects the mucous membranes. Exceptionally, adenopathies or metastases can be revealing. The treatment of choice is radical surgical excision with a safety margin of 3 cm of the healthy tissue around the tumor. Systematic lymphadenectomy is discussed. Nevertheless, it is necessary when the size of the tumor exceeds 2 cm and when the number of mitoses is greater than 10 per field or when it is a type with small cells or intermediate cells. Lymphadenectomy, on the other hand, is imperative in the presence of palpable adenopathies. Radiotherapy, as adjuvant therapy, is indicated in cervical-facial sites where large resection is not always possible and when the lymph node invasion is confirmed histologically [6, 7].

Surgical contraindications are logically related to radiotherapy or chemotherapy. In the case of metastatic dissemination, chemotherapy remains the therapeutic weapon of choice: the most widely used drugs are: vincristine, doxorubicin, cisplatinum, cyclophosphamide and etoposide [8, 6]. The evolution can be marked by the occurrence of metastases in half of the cases. The usual metastatic sites are: lymph nodes, liver, soft tissue and more rarely the lung and central nervous system. Approximately 35% of CNECs have a local recurrence within one year of treatment, especially when metastasis is present [9, 6].

Overall, the prognosis is poor and the survival rate at 3 years is 62% [6]. The factors that determine the prognosis are:

• age,
• larger than 2 cm,
• Histological criteria: a high mitotic index with a mitosis number greater than 10 per field, infiltration of lymphatic vessels, small cell or intermediate cell forms,
• the existence of adenopathies or metastases

**CONCLUSION:**

Primary cutaneous neuroendocrine carcinoma is a rare, aggressive cutaneous tumor whose incidence is increasing frankly, mainly due to the aging of the population. The diagnosis and therapeutic management of this tumor is not consensual. The localization at the level of lower limb remains very rare. The prognosis is bleak despite the current therapeutic progress

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