

# Merkel cell carcinoma: Report Of Two Patients

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## Abstract

Merkel cell carcinoma (MCC) is a rare malignant cutaneous tumor of the elderly that is characterized by an aggressive course with regional nodal involvement, distant metastases and a high rate of recurrence. It is a very rare tumor with an annual incidence of approximately 0.42 per 100,000. We describe two patients of Merkel cell carcinoma presented to us. Both patients were treated by surgery and chemotherapy. The response to treatment was very different in both the patients. The final outcome however has been very unsatisfactory in both patients.

## CASE 1

72 year old male presented with an ulcerated growth over the lower 1/3 of right leg. Examination revealed an 8x8 cm size ulcerated growth, with right leg edema and ipsilateral 9x8 cm fixed ilioinguinal nodes. An ultrasonography of the abdomen was suggestive of a large ilioinguinal mass on right side. A MCC was diagnosed. The rest of the metastatic workup was unremarkable. He was started on a combination of cisplatin and etoposide in a neoadjuvant fashion. After 4 cycles his primary site and nodal disease had disappeared and a wide excision with ilioinguinal node dissection was carried out. A pathological complete response confirmed by a synaptophysin stain was noted at both sites. However a small subcutaneous nodule over the anterior aspect of the middle third of the right leg was excised during surgery and was positive for MCC

Within 6 weeks of surgery, the patient developed a subcutaneous swelling over the dorsum of the right foot. A fine needle aspiration was positive for MCC. Multiple nodules appeared within one month over medial aspect of thigh and the inguinal region. He received three additional cycles of the same chemotherapy with a partial response, and was then lost to follow-up.

## Figure 1

Figure 1: Photographs of lesions before and after chemotherapy

### 1a. MCC primary site



**Figure 2**

**1b. Inguinal nodes**



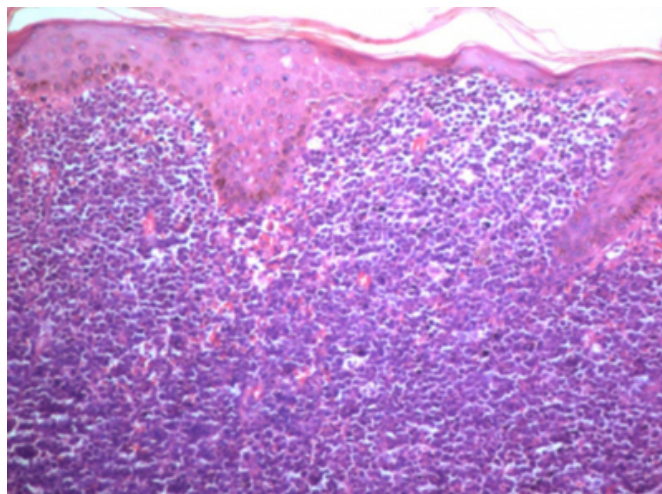
**Figure 3**

**1c. Complete response after chemotherapy**



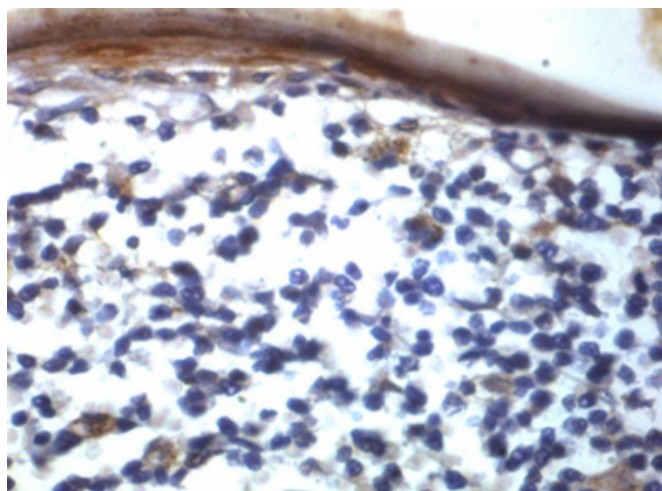
**Figure 4**

Figure 2: H & E slide showing diffuse sheets of small blue, ovoid cells with hyper-chromatic nuclei, fine chromatin and minimal cytoplasm. Mitoses were frequent and apoptotic cells were seen.



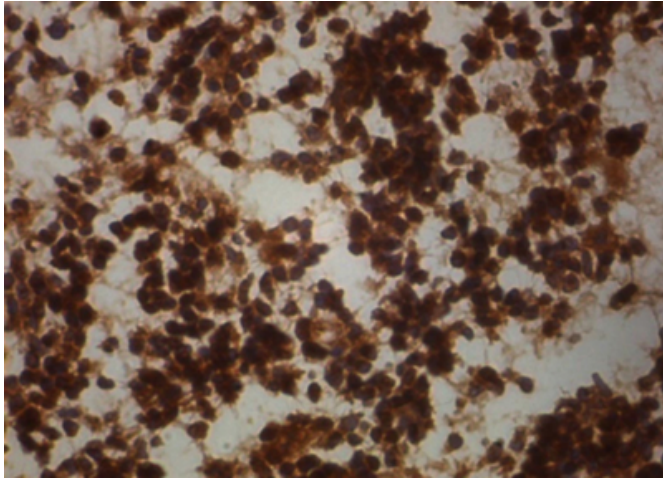
**Figure 5**

Figure 3: A focal positive reaction was observed for the pancytokeratin stain.



**Figure 6**

Figure 4: Synaptophysin stain is positive in all cells



**CASE 2**

A 65 year old man underwent an excision of a sebaceous cyst on the right upper back. The pathology was suggestive of MCC. He presented two weeks later with right supraclavicular and axillary lymphadenopathies. A fine needle aspiration confirmed the diagnosis of MCC. The metastatic workup showed multiple retroperitoneal lymphadenopathies with liver metastases. He was started on cisplatin and etoposide and had a partial response to chemotherapy. He received a total of 6 cycles. He later developed brain metastases within two months of completion of chemotherapy and eventually succumbed to the disease within 8 months of diagnosis.

**DISCUSSION**

Merkel cell carcinoma (MCC) is a rare cutaneous neuroendocrine tumor thought to be derived from a specialized epithelial cell, the Merkel cell. It is a nondendritic, nonkeratinizing, “clear” cell present in the basal cell layer of the epidermis, free in the dermis, and around hair follicles as the hair disk of Pinkus. MCC express both neuroendocrine (neuron-specific enolase, synaptophysin, chromogranin) and cytokeratin markers (cytokeratin 20, as a paranuclear dot, CAM 5.2) and is negative for S100 and the common leukocyte antigen ( $\text{CD}45$ ). The tumor may be solitary or multiple and occurs on the head and on the extremities. It involves the extremities in 40% of cases and the trunk in less than 10% of the cases ( $5,6,7,8,9$ ). On the face, the eyelids are involved more frequently ( $10$ ). There is a high rate of recurrence following excision, but, more important, it spreads to the regional lymph nodes in more than 50% of the patients and has a tendency to metastasize to the viscera and the central nervous system.

Ultraviolet radiation has been implicated as a factor in the development of MCC, due to the occurrence of the tumor on sun exposed skin ( $1,11$ ). Immunosuppression following organ transplantation is also associated with the development of MCC.

Recommended treatment for all stages of MCC is excision of the primary lesion with a 2 cm or greater margin whenever possible. All patients with no obvious lymph node disease should undergo sentinel lymph node biopsy ( $3,4$ ) at the time of wide surgical excision ( $5$ ). Radiotherapy is recommended to the site of primary lesion and to the draining lymph node basin in stage I and II disease. However, it is unclear that the combination of lymph node dissection and radiotherapy improves the chance of recovery over radiotherapy alone ( $12$ ).

Despite local excision, the incidence of local recurrence, regional lymph node metastases, and distant metastases is high and usually occurs within 2 years of primary diagnosis. Adjuvant chemotherapy should therefore be considered as part of the initial management. Metastatic disease should be treated with chemotherapy, followed when feasible by surgical resection or consolidation radiotherapy if a response is obtained with chemotherapy. The chemotherapy is based on regimens used for small-cell lung cancer, mainly combinations of cisplatin or carboplatin and etoposide. Other chemotherapy agents to consider are topotecan, doxorubicin, cyclophosphamide, and vincristine.

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