

Giant Merkel cell carcinoma of the left arm.

Case report



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Giant merkel cell carcinoma of the left arm. Case report.

Merkel cell carcinoma is a rare tumor of dermal origin generally found in sun exposed skin.

We report the case of a woman of 76 years old presenting a large vascularized Merkel cell carcinoma (MCC) of the left arm lateral to the elbow joint, infiltrating the musculo-fascial plane who was treated with surgical therapy and post operative radiotherapy.

KEY WORDS: Merkel cell carcinoma, Neuroendocrine tumor, Surgery.

Case report

We report the case of a woman of 76 years old who arrived to our Institute presenting a large vascularized lesion of the left arm lateral to the elbow joint, infiltrating the musculo-fascial plane who was treated with surgical therapy. The mass was painless and the ipsilateral axillary lymph nodes were negatives.

Physical examination showed a large ulcerated cutaneous tumor measuring 13 cm by 10 cm on the left arm near the elbow .

Imaging of the primary tumor and regional lymph nodes was done with CT and MRI (Fig. 1) that showed a large mass with regular margins infiltrating the lateral group muscles. No distal metastasis were evaluated using TC. According to the staging proposed by Yiengpruksawan it is classified as IB stage. Using TNM classification is T4N0M0; stage III in according to the American Joint Committee on Cancer Staging System (2002) (Tab. I).

The diagnosis was confirmed by a biopsy of the mass. Histologically the tumour was composed by large solid cellular masses infiltrating the dermis and subcutis, sparing the epidermis (Fig. 2). The neoplastic nests were composed by uniform, small, round to oval cells with scant cytoplasm and vesicular nuclei containing small nucleoli. Numerous mitotic figures and apoptotic bodies were visible. Invasion of lymphatic vessels by tumour cells was a feature (Fig. 3).

Immunohistochemically tumour cells showed typical perinuclear dot-like positivity to low molecular weight cytokeratin and more diffuse positivity to neuroendocrine markers (synaptophysin, neuron-specific enolase, PGP9.5 and focally chromogranin-A) (Fig. 4). The differential diagnoses on morphologic grounds alone included a lymphomatous process and a metastatic small cell carcinoma, these entities could be excluded by means of the immunophenotype of tumour cells and by accurate clinical evaluation of the patient.

The patient, according to the literature, was subjected to a wide surgical excision (margins 2-3 cm free from tumor) of the lesion (fig. 5). Plastic surgical methods of lateral skin flap of rotation and dermo-epidermal thick-split graft from inguino-crural area of the left thigh was used to cover the deficit area.

Post operative radiotherapy was done and the prescribed dose was 50 Gy.

Patient died after 6 months for metastatic dissemination .

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TAB. I

Primary tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- Tis Carcinoma in situ
- T1 Tumor 2 cm or less in greatest dimension
- T2 Tumor more than 2 cm, but not more than 5 cm, in greatest dimension
- T3 Tumor more than 5 cm in greatest dimension
- T4 Tumor invades deep extradermal structures (i.e. cartilage, skeletal muscle, or bone)

Stage Grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
	T3	N0	M0
Stage III	T4	N0	M0
	Any T	N1	M0
Stage IV	AnyT	Any N	M1

Discussion

Merkel cell carcinoma is a rare tumor of dermal origin generally found in sun exposed skin.

The annual incidence is estimated to be 0.42 per 100,000 from the Connecticut Tumor Registry database from 1990 to 1997.¹

Toker first² described Merkel Cell carcinoma (MCC), which he called trabecular carcinoma of the skin, in 1972. The cell of the origin was described as an epi-



Fig. 1: NMR showing the large mass of the left arm infiltrating musculo-fascial plane.

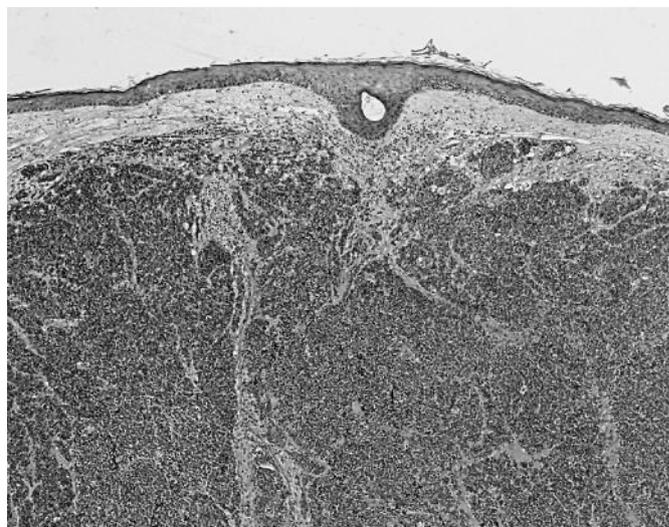


Fig. 2: Large basophilic cellular aggregates infiltrating the dermis and sparing the epidermis. (orig. magn. 40x).

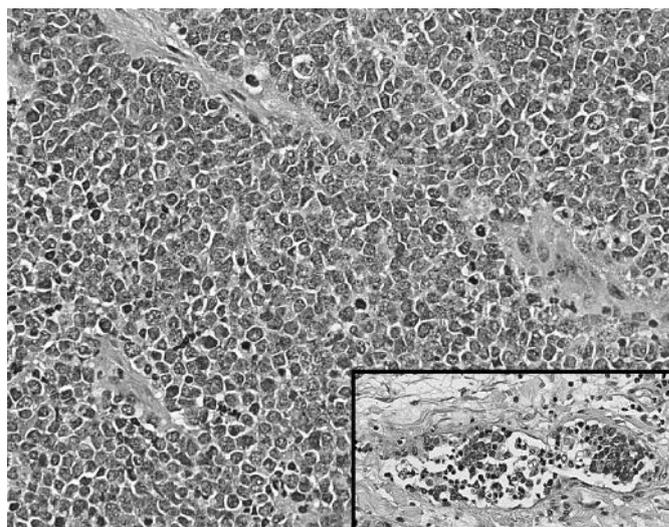


Fig. 3: Higher magnification showing cellular details. Note the uniformity of cells with scant cytoplasm and vesicular nuclei and the numerous mitotic figures. In the insert, a lymphatic vessels invaded by tumour cells. (orig. magn. 200x).

dermal, nondendritic, non-keratinocytic cell that he referred to as a tactile cell.³ These cells are of neural origin and found within the basal layer of the epidermis either alone or in groups.

Since its initial description, there has been considerable debate with regard to its origin and treatment. Merkel cell carcinoma is the most common name for this skin cancer because of the presence of neurosecretory granules within tumor cells.⁴

MCC usually presents as a rapidly growing, painless nodule of 2 cm average diameter with overlying, skin discoloration ranging from pink to violet⁵⁻⁸.

The median age at the presentation is 64-68 years. Both

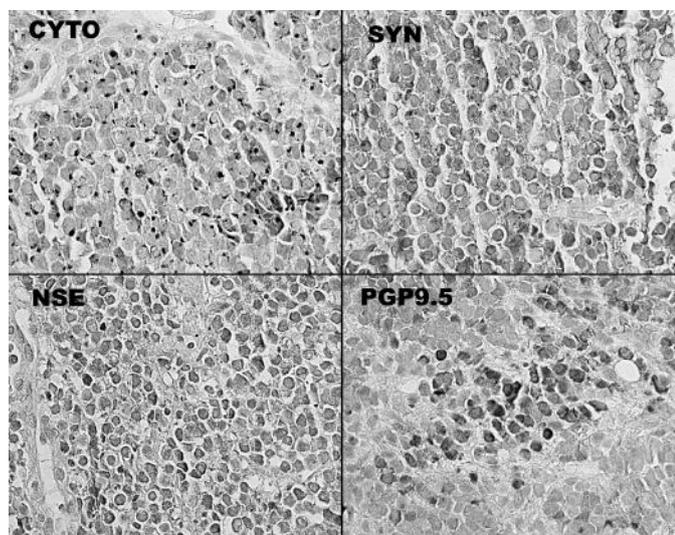


Fig. 4: Immunohistochemical results: dot-like perinuclear positivity to low molecular weight cytokeratin (CYTO), diffuse positivity to neuroendocrine markers: synaptophysin (SYN), neuron-specific enolase (NSE) and PGP9.5. (orig. magn. 400x).



Fig. 5: Large vascularized lesion of the left arm lateral to the elbow joint.

sexes are affected but there seems to be a slight male predominance⁹⁻¹³.

Ultraviolet (UV) radiation seems to be a significant etiologic factor given that 85% of lesions appear in sun-exposed sites and the Caucasians with fair skin have the highest incidence of disease.

Occurrence of this tumor is also reported after radiation¹⁴⁻¹⁶ and immunosuppressive therapy¹⁷ or in association with other malignancies.

It has been associated with squamous cell carcinoma, basal cell carcinoma of skin; haematological malignancies and with breast and ovarian adenocarcinomas.¹⁸⁻¹⁹

Most commonly involves the head and neck (50% to 55%) followed by the extremities (38% to 40%) and trunk (5% to 13%)⁵⁻²⁰.

This neoplasm is characterized by a high incidence of local recurrence (12% to 50%), regional nodal metastasis (17% to 76%) and distant metastasis (12% to 59%); it also has a high mortality rate (20% to 55%).^{16, 21-26} Reported 2-year and 5-year survival rates are 72% and 30% to 60% respectively²⁸⁻³⁰.

All the patients are staged as proposed by Yiengpruksawan et al²⁶ at their initial presentation according to the absence (stage I) or presence (stage II) of positive lymph nodes within the draining nodal basin and by the presence of systemic metastases (stage III). For stage IA the tumor is smaller than 2 cm, stage IB 2 cm or larger.

Stage I disease, for disease that remains localised to the primary site, the mainstays of treatment are surgery and radiation. 5-year survival has been quoted at 64%³³. The addition of radiotherapy reduced the local failure from 39% to 26% and the regional failure from 46% to 22%³¹.

On immunohistochemical studies, MCC express both epithelial and neuroendocrine markers³²⁻³³. Cytocheratin 20 is a sensitive and specific marker for MCC and is helpful in distinguishing between MCC and other malignant and benign neoplasm³⁴⁻³⁵.

The two potentially curative treatment modalities for patients with MCC are surgery and radiotherapy³⁶, that can obtain a loco-regional control.

Conclusion

MCC is an extremely rare cutaneous neoplasm. Stage I disease should be treated with surgical excision, using either wide local excision with 2 to 3 cm margins, dissecting to fascia and may be followed by lymph node dissection. Postoperative radiation may also be considered to improve locoregional control.

Treatment options may be controversial, but its clear that this tumor is very aggressive, and often has a poor prognosis.

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