

Merkel cell carcinoma of the leg.

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Section: Musculoskeletal system

Imaging Technique: MR

Case Type: Clinical Cases

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Patient: 60 years, female

Clinical History:

A 60 year old woman referred for nodular lesions on the leg

Imaging Findings:

A 60 year old woman was referred to our institution for nodular violaceous lesions on the lateral aspect of her right leg with surrounding erythematous reaction. A doppler ultrasound examination performed ruled out the presence of thrombophlebitis and she was then treated for a presumed erysipela without noticeable improvement. A biopsy done revealed the diagnosis of a neuroectodermal tumor. No research for a sentinel lymph node was performed because of the inflammatory and ulcerative characteristics of the tumor. An extensive work-up done, ruled out the presence of distant metastasis including lymph nodes. Treatment consisted on isolated limb perfusion with Melphalan and the subsequent clinical follow-up was pertinent for a local control of the disease but associated with a progressive necrosis of the cutaneous lesion necessitating intensive local treatment.

Discussion:

Merkel cell carcinoma (MCC) is a rare highly malignant primary neuro-endocrine tumor of the skin first described by Toker in 1972 and termed « trabecular carcinoma of the skin » based on its histological characteristics(1-3). But the presence of neurosecretory granules commonly found in Merkel's neuroendocrine cells of the basal layer of the epidermis explains the other variety of names under which the lesion was reported including « primary neuroendocrine carcinoma of the skin » and « Merkel cell carcinoma »(1-3). It is still controversial whether this tumor arises from Merkel cell or from a pluri-potential cell in the epidermis with the majority favoring the latter hypothesis(1,3). MCC is a tumor of the elderly with an age range between 35 and 97 years and a mean age of 70 years(1,2,4) with a near equal male /female ratio(1) or a slight female predominance(2). The most common sites of occurrence are the sun-exposed areas of the skin(1,4) with the head and neck region accounting for 55 % of cases and the extremities 40% with a predilection for the lower limb. This anatomic distribution as well as the occurrence of basal and squamous carcinomas in close proximity suggested a common carcinogenic stimulus such as UV light(1,4) ;however the documentation of other rare cases on unexposed areas of the skin(1,3) such as the trunk suggests a more complex pathogenesis. On routine HE stains the tumor is made of round cell with high number of mitoses and necrotic cells(2). 3 types of MCC have been recognized histologically : 1: Trabecular type; 2: Small cell type similar to neuroendocrine carcinoma of the lung. 3:Intermediate type which is the most frequent. Differential diagnosis includes(5) melanomas, spindle cell hemangioendotheliomas, localized limb metastases of epithelioid sarcomas, clear cell sarcomas, malignant peripheral nerve sheath tumors, porocarcinomas. Usually patients with MCC present clinically with an erythematous or violaceous painless skin nodule arising on sun-exposed skin(1,2) mimicking benign skin lesions as well as other skin neoplasms such as basal cell carcinoma(1,2,4). The aggressiveness of this tumor is expressed by the high rate of local recurrence and by the lymphatic and distant metastases(1-2-4). Regional lymph node metastases occur in 30% at initial presentation (1,2) and subsequently

during the course of the disease in 55-83% of subjects(2) whereas hematogenous spread occurs in up to 50% within two years(1) and is unusual initially occurring with decreased frequency to the liver, bone, brain, lung and skin. Local recurrence was reported in 25-77% of cases within 4 to 8 months after excision (2) this high rate is explained by positive margins upon surgery, and the presence of an extensive lymphatic system in the head and neck(4). The primary treatment of choice to avoid recurrence is wide surgical excision with 1 to 3 cm wide excision as is recommended for melanoma (4). Localized hyperthermic limb perfusion with chemotherapeutic agents like Melphalan and Cisplatin is used when the lesion is localized to an extremity. Limb amputation is only limited if there is no response to limb perfusion or immunotherapy. MCC unlike melanomas are radiosensitive and radiotherapy is recommended as adjuvant therapy if no wide excision is possible. Imaging modalities are important initially as well as in the follow up .At CT the primary skin lesions as well as the distant metastases and lymph nodes appear hyper or isodense to muscles. The presence of streaky linear and reticular densities in subcutaneous fat surrounding the primary lesion represents lymphatic invasion especially if associated with regional lymph nodes(1,2).CT Scan is also indicated initially to detect pelvic and retro-peritoneal lymph node and to exclude the diagnosis of metastatic small cell carcinoma of the lung that shares histologic findings with MCC(4). At MRI the signal features of the primary lesion are non specific with a long T1 and T2. However, the association i with an adenopathy , a satellite nodule should raise the suspicion of Merkel cell tumor(1).The peri-tumoral streaky high signal in peri-tumoral fat indicates, as on CT, local lymphangitic invasion thereby delineating the boundary of local tumor spread which is important for surgical resection.(1). The imaging modalities are important to define the extent of the tumor ,however the definitive diagnosis requires the use of immunohistochemistry allowing the isolation of specific tissue markers such as NSE which are positive in the majority of cases and low molecular weight CAM5,2 which are considered pathognomonic for MCC as well as the peri nuclear dot cytokeratin staining pattern(2-5).

Differential Diagnosis List: Merkel cell carcinoma of the leg

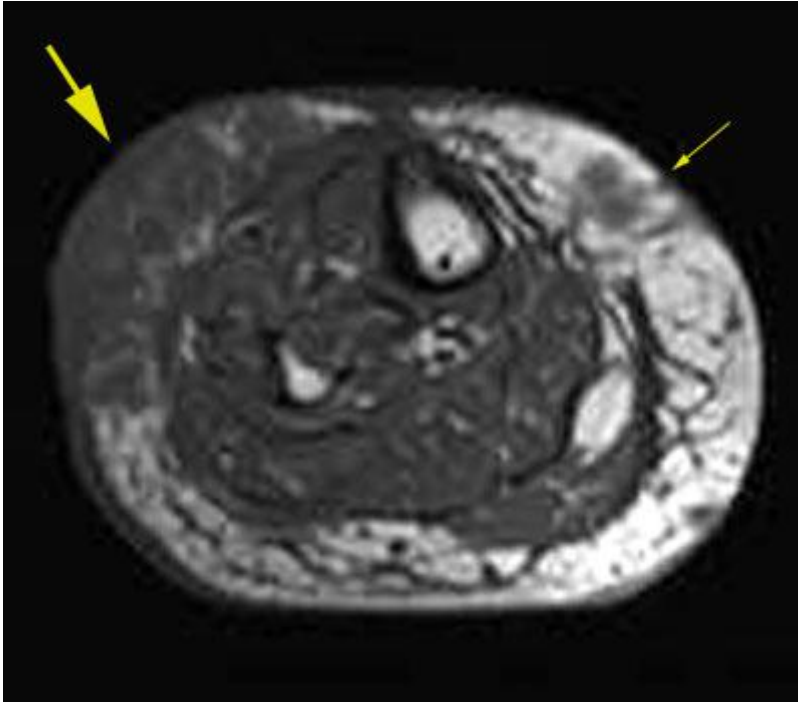
Final Diagnosis: Merkel cell carcinoma of the leg

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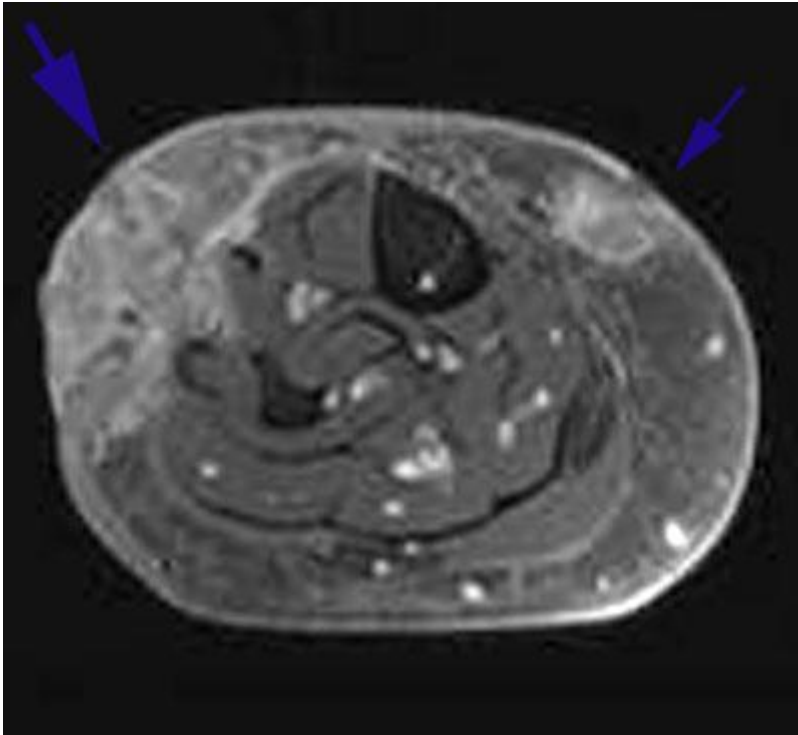
Figure 1

a



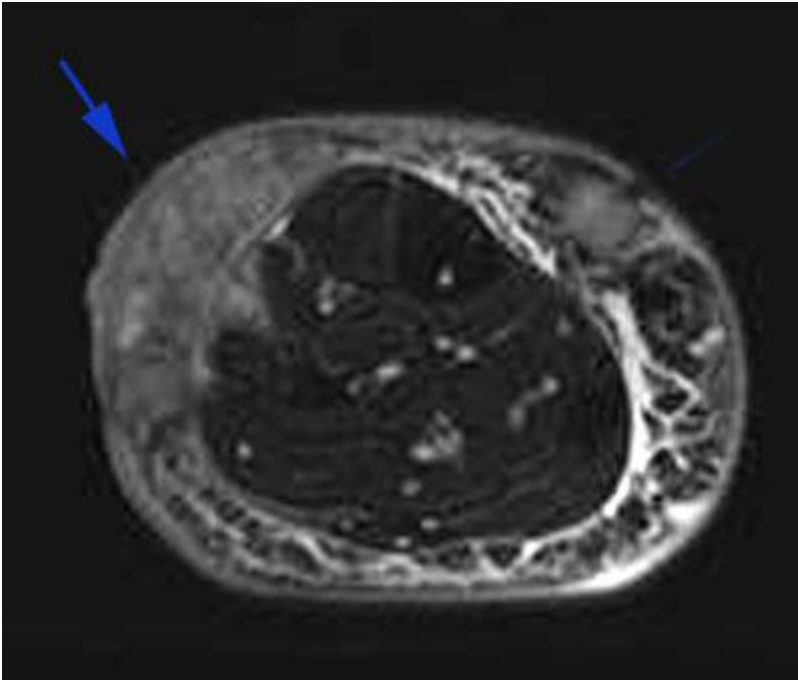
Description: Axial T1 - weighted SE image showing the subcutaneous lesion (large arrow) of decreased signal intensity , well-demarcated from the high signal of the surrounding subcutaneous fat. Note the distant antero-medial nodule (small arrow). **Origin:**

b



Description: Axial T1 -weighted SE with Gadolinium showing both the enhancing main tumor (large arrow) as well as the nodule (small arrow). **Origin:**

c



Description: On the axial T2 FSE with fat saturation the lesion (arrow) appears with intermediate signal intensity **Origin:**

d



Description: Coronal T1 -weighted SE with gadolinium showing the vertical extension of the lesion (arrow). **Origin:**