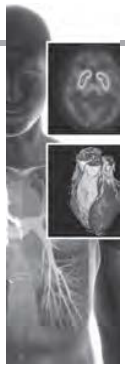

Diagnostic Imaging in Patients with Merkel Cell Carcinoma

Merkel Cell Carcinoma (MCC) is a rare and invasive cutaneous neuroendocrine tumor with a mortality rate of about 25%. Accurate assessment of lymph node involvement in patients with MCC significantly predicts overall outcome. Due to the rarity of this very dangerous disease, only a few imaging reports on MCC have been published and thus, to date, there is no accepted imaging algorithm for MCC. For the initial stage of MCC, general recommendations include ultrasound, chest X-ray, and MRI, but recent papers suggest an increasing use of the focal node and FDG-PET/PET-CT become important. Merkel cell carcinoma is a rare and very aggressive neuroendocrine tumor of the skin. It mainly develops on sun-exposed head and neck areas. In 1972, Toker described an unknown trabecular skin carcinoma in 5 Caucasian patients. In 1982, Tang and Toker proposed that MCC originates from Merkel cells, a hair follicle receptor mechanism.

KEYWORDS: Diagnostic Imaging • Medical Imaging • Patients • Merkel Cell Carcinoma

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Introduction

MCC usually grows rapidly and presents as a firm, non-soft, dome-shaped red, purple or purple nodule. The outer layer of skin is smooth and shiny, sometimes with features of ulcers, acne or varicose veins. MCC tends to metastasize to regional lymph nodes and in 50% of patients it spreads hematogenously to other organs, i.e. liver, bone, brain, and lung. In 1993, Haag and colleagues identified a commonly used staging system: stage I is defined by local disease without lymph node involvement or metastasis. Distant, stage II carcinoma has spread to lymph nodes but no systemic metastasis has been detected, and in stage III, distant metastasis can be detected.

Diagnosis of MCC can be difficult because in many cases MCC lesions mimic benign skin lesions. Unfortunately, in clinical practice, highly suspicious lesions of Merkel cell carcinoma are often incorrectly biopsied or resected with narrow margins. In fact, patients with indistinct new skin lesions should be examined clinically, and lesions that are still highly suspicious of Merkel cell carcinoma should be resected with clear and broad margins. The diagnosis and management of lymph node metastases in patients without a primary tumor can be difficult. In particular, metastasis from MCC can mimic metastasis from other small cell tumors, eg lung carcinoma. In 2-19% of patients, no primary tumor was found, defined as unspecified primary MCC (MCUP). Due to the rarity of this disease, the literature on MCCUP is very limited [1, 2].

Agelli performed several epidemiological studies showing that between 1986 and 2001, the incidence of age-matched MCC tripled with an annual increase of 8%. This increasing incidence has contributed significantly to the growing interest in the management of patients with Merkel cell carcinoma. Unfortunately, there is little literature on imaging algorithms in Merkel cell carcinoma patients and no widely accepted guidelines for Merkel cell carcinoma imaging. This article reviews the imaging literature on Merkel cell carcinoma discussing the role of the latest imaging and diagnostic tools [3].

Discussion

Ultrasound is a very accurate and economical technique for tumor classification. For Merkel cell carcinoma, evaluation and staging of the neck should begin with ultrasound examination. Primary skin lesions may present as single or multicentric hypoechoic solid nodules arising from the dermis and extending into the subcutaneous fat layer, with varying degrees of posterior acoustic transmission. The ultra sonographic features of Merkel cell carcinoma appear similar to more common skin tumors such as melanoma or basal cell carcinoma. It has been shown that in areas easily accessible by ultrasound, such as the neck, it is possible to distinguish malignant from benign nodes with an accuracy of 89% to 94%. In addition, ultrasound has an important role in real-time imaging during fine-needle biopsies of non-palpable Merkel cell carcinomas. Except for a few published case reports, ultrasound-guided

and unguided fine-needle aspiration biopsies have rarely been described in patients with MCC. Definitive diagnosis of metastatic disease is difficult with fine needle aspiration cytology alone. The cytology resembles many other malignancies such as malignant lymphoma and melanoma. However, the MCC FNA can provide an accurate and reliable diagnosis of primary or recurrent metastatic lesions. In patients with positive lymph nodes, full-body imaging is recommended to detect distant metastases [4].

Sentinel Node Biopsy offers the unique ability to detect metastases and micro-metastasis and subsequently lymph node drainage of lymphatic metastases in patients with melanoma, squamous cell carcinoma and MCC using lymphatic tomography. SLNB in patients with Merkel cell carcinoma appears to be a reliable staging technique, while the prognosis regarding positive ganglion cystic tumor status remains unclear. In up to two-thirds of patients with stage I MCC, regional lymph node enlargement is diagnosed at initial presentation of SNLB, and only 7% to 31% of lymph nodes are clinically palpable in patients with stage II disease [5].

Lymphatic pathways in the head and neck region vary more than anywhere else in the body and are difficult to predict with precision. Occasionally, head and neck lymph node scintigraphy fails to identify a clear pattern of lymphatic drainage. In particular, unexpected lymph node drainage is observed in 37% to 84% of cases and is often missed without radiographic guidance. A negative focus biopsy appears to be a suitable prognostic factor for disease-free survival. Thus, false-negative lymph node dissection results in incomplete MCC staging and positive but unnecessary complete lymph node dissection in patients with true stage I disease. However, they have shown that in 5-6.8 % of patients with melanoma or Merkel cell carcinoma of the head and neck region, no detectable lymphadenopathy. In particular, in 5 of the 74 clinical lymph node patients who underwent preoperative lymphangiography, lymphatic tomography did not identify positive lymph node metastases. In 2002, Nguyen and colleagues proposed lymphatic scintigraphy combined with perioperative lymphatic mapping [6].

Conclusion

▪ Computed Tomography (CT)

Because of the usefulness of CT in imaging head and neck lymph nodes as well as subcutaneous fat and visceral metastases, several effects Authors propose that CT is a reliable imaging modality for the initial staging of patients with Merkel cell carcinoma. In particular, they have suggested a sensitivity and specificity ratio of 47% and 97%, respectively, with a positive and negative predictive value of 94% and 68% for the diagnosis of lymph node involvement, respectively. CT scan However, Peloschek and colleagues confirmed a specificity of 96.2% and sensitivity of 89.1% for computed tomography in the imaging of Merkel cell carcinoma, including involvement of lymph nodes as well as evaluate for distant metastases [7, 8].

Compared with muscle, primary cutaneous lesions present as uniformly dense to slightly dense round skin nodules extending subcutaneously. A cutaneous fat mass adjacent to the primary lesion suggests tension and edema due to lymphatic invasion. In addition, enhanced CT scans can show high-density enlarged lymph nodes and soft CT scans can show high-density tissue nodules, often without clinical signs, indicating a high degree of attenuation focal length of metastases. Swollen lymph nodes are mainly in the neck, especially the parotid region, then in the armpit, mediastinum, retro peritoneum, and groin. Distant metastases include local and retroperitoneal lymph nodes, liver, bone, brain, and lung. Using computed tomography, intra-abdominal metastases present as proliferative vascular lesions with annular enhancement. Soft tissue metastases may involve the chest wall or the abdomen with skeletal muscle invasion. Gollub and colleagues conducted a study of 12 patients with MCC and showed the ability of CT to detect visceral and lymph node metastases. They recommend follow-up CT scans at 3, 6, 12 and 18 months after initial therapy to detect recurrence [9, 10].

Acknowledgement

None

Conflict of Interest

None

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