MERKEL CELL CARCINOMA OF THE FACE: TWO CASE REPORTS AND LITERATURE REVIEW

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SUMMARY – Merkel cell carcinoma is an aggressive tumor of the skin deriving from neuroendocrine cells with unpredictable clinical behavior and poor clinical outcome. Merkel cell carcinoma is relatively rare with some 600 cases reported, but seemingly with an increasing incidence due to the increased sun exposure and immunosuppression. There are increasing reports of Merkel cell carcinoma occurring in transplant patients receiving immunosuppressive therapy. We present two cases of Merkel cell carcinoma of the facial skin, one of them in a patient who received immunosuppressive therapy. According to the literature, standard therapy for Merkel cell carcinoma includes large scale surgical excision of the primary tumor, neck dissection for palpable nodes, and in most cases subsequent radiotherapy. This article also reviews some recent reports of Merkel cell carcinoma regarding the diagnosis, clinical course and therapeutic modalities.

Key words: *Carcinoma – Merkel cell, etiology; Carcinoma – Merkel cell, therapy; Skin neoplasms, therapy; Organ transplantation, therapy*

Introduction

Merkel cell carcinoma (MCC), also named trabecular carcinoma, cutaneous small cell carcinoma, or undifferentiated carcinoma, is an uncommon aggressive form of skin cancer. Toker was the first to describe this tumor in 1972[1], and some 600 cases have since been reported. MCC has several characteristics similar to malignant melanoma such as unpredictable biologic behavior, early regional lymph node involvement, early distant metastases, high locoregional recurrence rate[2], and high mortality[3]. This tumor mostly occurs in elderly whites, particularly on sun exposed skin of the head and neck. The reported annual age-adjusted incidence of MCC is 0.23 for whites and 0.01 for blacks *per* 100 000[4]. MCC has been described in patients from 15 to 97 years of age, but predominantly between the sixth and seventh decade of life[3,5,6]. There is a slight female predominance[6,7], except for oral MCC[5,8]. An epidemiologic study in the United States showed an increased regional incidence rate correlating with the increasing sun exposure as measured by UVB solar index[4].

MCC arises from neuroendocrine cells with features of epithelial differentiation[9], in the dermis and subcutaneous tissue[10]. Merkel cells are round cells found in the basal layers of the epidermis, in the dermis, nails and oral mucosa in humans[11,12]. They commonly surround tactile hair follicles in organized innervated clusters and function as neurosensory transmitters for touch reception on the skin[13,14].

Clinical lesion is a painless, firm, raised nodule with a red, pink or bluish color and diffuse margins[5]. Epidermis is usually intact, but more advanced lesions become ulcerated[5]. Patients often present with a history of a slow grow-

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ing, pink or violaceous plaque or subcutaneous nodular lesion that suddenly and rapidly increases in size[5]. Up to 45% of patients have lymph node involvement at presentation[6,15].

The head and neck area is the most common site of tumor occurrence, and according to literature 35%-53% of MCC arise in this region[3-6,16], 7%-30% on lower limbs[5,16], 15%-32% on upper limbs[5,16], and 4%-26% on the trunk[5,6,16]. Rice *et al.* analyzed the incidence of MCC in the head and neck, and found cheeks to be the most common primary site (29%), followed by eyelids (18%), forehead (17%), lips (9%), ears (7%), nose (5.4%), scalp (4%) and chin (2%)[17].

The tumor occurs more frequently in patients with immunodeficiency disorders. According to literature, 53 cases have been reported in transplant recipients[4,15,18]; 18 cases in B-cell lymphoma patients[4]; 2 cases in patients with previous diagnosis of chronic lymphocytic leukemia[15]; 1 case in a patient with Recklinghausen neurofibromatosis[19], and 1 case of primary nodal MCC in a patient with HIV infection[20]. In the recent literature[21,22] there are reports of MCC arising after therapeutic immunosuppression.

The clinical and histopathologic differential diagnoses include basal cell carcinoma[3], melanoma, metastatic oat cell carcinoma, adnexal carcinoma, lymphoma, extraskeletal Ewing sarcoma and neuroblastoma[5,9,13]. A thorough clinical evaluation, light and electron microscopy, and a defined panel of immunohistochemical studies[5] best accomplish the diagnosis. Histologically small round or oval tumor cells form dense cohesive sheets with a variable trabecular pattern, having hyperchromatic nuclei and a high rate of mitosis[5]. Because of the highly variable and undifferentiated appearance under light microscopy, one study showed misdiagnosis in 66% of lesions examined by light microscopy alone[13]. MCC ultrastructural characteristics include 75 to 200 nm, electron-dense, neurosecretory granules, perinuclear whorls of intermediate filaments, and primitive nondesmosomal junctional complexes[5]. Immunohistochemically tumor cells consistently express cytokeratin 8 and cytokeratin 20, and variably express neuron-specific enolase, epithelial membrane antigen and chromogranin[5]. The expression of cytokeratin 8 is common in many carcinomas[5]. Cytokeratin 20 positivity is predominantly seen in Merkel cell and salivary gland small cell carcinomas[5], and is usually negative (67%-97%) in lung small cell neoplasms[23,24]. A combination of thyroid transcription factor-1 (sensitive and specific marker for small cell carcinomas of the lung) and cytokeratin 20 has been suggested to assist in the differentiation of metastatic small cell carcinoma of the lung from Merkel cell carcinoma[24].

Since MCC has some characteristics similar to malignant melanoma, and some similar to squamos cell carcinoma (SCC), there is no consensus regarding tumor staging (such as TNM) for primary lesions. Most of the tumors described in the literature were not staged, with the exception of the article from Memorial Sloan-Kettering hospital[25]. The authors have classified the disease into three stages: stage I – local disease, stage II – locoregional disease, and stage III – distant metastases. Stage I was further divided into IA and IB according to primary lesions smaller and larger than 2 cm, respectively.

The management of MCC has followed the principles of malignant melanoma or SCC treatment. Since no large patient series with MCC have been reported, there is no general consensus regarding optimal treatment. The primary method of treatment is surgery. After standard surgical excision MCC tends to recur locally and develop regional nodal spread[26]. The incidence of locoregional recurrence is 40%-65 % and 40%-82% if patients develop metastatic disease[3,10,27].

Primary lesions should be treated with wide excision with 3- to 5-cm margins[3,5,7,28,29] or Mohs micrographic surgery[26,28,30]. In one study, lesions treated with standard surgical excision were associated with 31.7% of local persistence and 48.8% of regional metastasis. In contrast, lesions treated with Mohs micrographic surgery were associated with 8.3% of local persistence and 33.3% of regional metastasis[26]. Therapeutic regional node dissection should be performed for palpable lymphadenopathy[5]. However, for lesions located in areas of predictable nodal drainage, prophylactic regional node dissection is recommended[3,5,29]. According to the literature, elective lymph node dissection decreased the rate of locoregional recurrence[5,25,31], but it was not associated with improved overall survival[25,31]. In one study, the incidence of micrometastases in patients undergoing prophylactic lymph node dissection was 100%[31]. Some authors suggest sentinel lymph node biopsy in evaluating occult lymph node involvement, thus limiting the potentially unnecessary morbidity of more comprehensive lymph node dissections in patients who do not yet have metastatic involvement[2,30,32,33].

Literature data support the use of combined treatment with surgery and radiation therapy for patients with advanced locoregional MCC[5,13,29,34]. Postoperative radiation therapy to primary site is recommended in addition if there is evidence of incomplete excision[5,35], or primary lesion is larger than 1.5 cm or is resected with narrow margins (<2 mm) or shows histologic evidence of lymphatic invasion[5,13]. Postoperative radiation therapy to both primary site and

regional lymph node areas is recommended if histologically positive nodes are found[5,13,29]. External beam radiation therapy is highly effective when given as consolidation after surgery with no locoregional recurrences according to two studies[6,36]; with 9% in-field recurrence according to one study[34], or 79% achievement of locoregional control according to another one[3]. In contrast, excision and irradiation of relapsed disease yielded an in-field recurrence in 42% of irradiated patients[36]. Some authors suggest that radiotherapy after Mohs surgery reduced persistent metastases in transit and nodal disease[25]. Local excision together with regional lymph node clearance and adjuvant radiation treatment is associated with an improved survival[3].

Chemotherapy has been used alone or in combination with radiation therapy for advanced or disseminated disease in patients with good performance status[5,34,37]. The role of chemotherapy in the treatment of MCC is unclear. MCC is chemosensitive but rarely chemocurable in patients with metastasis or locally advanced tumors[38]. A high incidence of toxic death (3.4%-7.7%) due to chemotherapy is reported in the literature[37,38]. Cyclophosphamide/doxorubicin (or epirubicin)/vincristine combination +/- prednisone was the most commonly used chemotherapy regimen, with an overall response rate of 75.7%[37]. Etoposide/cisplatin (or carboplatin) was the next commonly used, with an overall response rate of 60%[37].

After developing systemic disease, patients receiving no treatment survived for an average of 3 to 4 months[5]. Radiation therapy has been effective in palliating symptoms resulting from systemic disease, but has had little effect in the prolongation of life. The median survival for patients receiving radiation, chemotherapy or both was less than 8 months[7].

Case Reports

Case 1

E. K., a female Caucasian born in 1937, nonsmoker with unremarkable history until 1996 when she developed dermatomyositis affecting several joints and skin. She started immunosuppressive therapy (azathioprine (Imurek, GSK) 3x50 mg and prednisolone (Aprednisolon, Merck) 1x25 mg) and received it for two years.

In June 1998 she noticed a change on her left cheek, in the form of slightly elevated induration, 2 cm in diameter, moderately darker than the surrounding skin, and with small telangiectatic vessels on its surface. The tumor borders were unclear. In November 1998, a biopsy revealed MCC. Preoperative tests included positron emission tomography (PET), computed tomography (CT) of the head and neck, and ultrasound (US) of the neck and abdomen. PET showed a 'hot spot' on the left cheek, 17 mm in diameter. None of these examinations showed any sign of metastatic disease.

In January 1999 the tumor was excised with wide margins (the specimen measured 53 mm in diameter) and the resected specimen included lateral lobe of the parotid gland. The branches of facial nerve were preserved. The operation included selective supraomohyoid neck dissection on the left side. The defect was reconstructed with free radial forearm flap anastomosed to facial artery and retromandibular vein.

The pathologic diagnosis was MCC. The tumor measured 17 mm in diameter and 16 mm in thickness. Underlying muscles were not invaded by the tumor, and lateral free margins were at least 18 mm wide. Eighteen lymph nodes were isolated from the neck dissection specimen, without evidence of tumor. Pathologic finding was staged as pT1 pN0.

The patient refused recommended adjuvant therapy. In June 1999 she developed 2 regional neck metastases in regions I and II, which were removed by selective dissection. Again, she refused additional therapy. In December 1999 additional dermal spots of MCC occurred in the head and neck area. CT scan of the lung showed small multiple metastases. In February 2000 cervical metastases increased

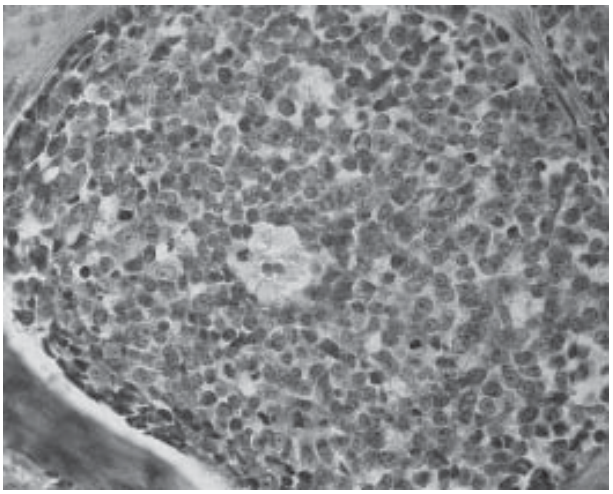

Fig. 1. Hematoxylin-eosin stained histology section of MCC specimen (X400 magnification). Tumor cells have round hyperchromatic nuclei and scarce cytoplasm. There are pseudorosette-like formations and numerous mitoses.

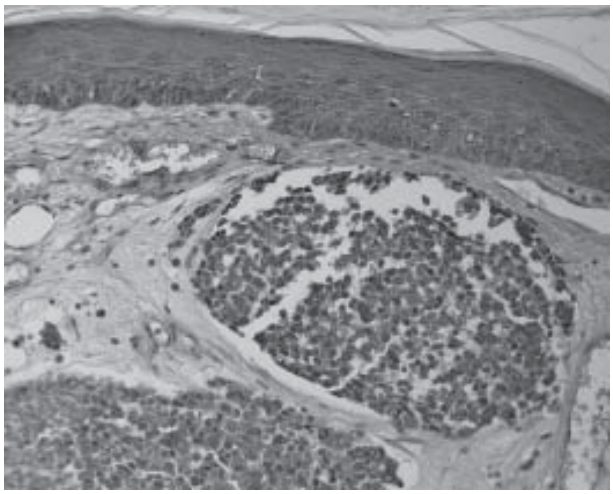

Fig. 2. The same specimen under X200 magnification. Immunohistochemical reaction positive for cytokeratin.

and the patient had to be tracheotomized, whereafter she refused any kind of therapy and was transferred to a hospice, where she died in March 2000.

Case 2

S. J., a male Caucasian born in 1920, nonsmoker, with unremarkable history, treated only with antihypertensive therapy for several years (diuretics and beta blockers). In December 1999 he noticed a small skin change of 5 mm in diameter on his right cheek. The change was removed by a dermatologist in January 2000, and the pathologic diagnosis was MCC (Figs. 1 and 2). In February 2000 he noticed node enlargement in the right submandibular region. Fine needle aspiration biopsy indicated metastatic carcinoma, and the patient was referred to an ENT department. US examination revealed nodal enlargement in neck regions I to IV, without invasion of blood vessels. Abdominal US and X-ray of the thorax showed no signs of distant metastases. The patient was operated on in May 2000 at ENT Department in Zagreb. Postoperative scar on the cheek was excised in full thickness and with 1-cm lateral borders. Ipsilateral radical neck dissection was performed with preservation of the eleventh nerve.

Histologic analysis found 24 lymph nodes in the resected specimen, 10 of them invaded by MCC in regions I to IV. Two nodes in region II had extranodal spread of tumor. The patient received postoperative adjuvant radiotherapy (6000 cGy in 30 fractions). Twenty-two months of initial diagnosis and sixteen months of therapy completion he is alive and without evidence of the disease.

Discussion

MCC is an aggressive tumor with unpredictable clinical course which can be compared with malignant melanoma. The initial presentation of MCC is often unremarkable, leading to delay in diagnosis and treatment. The reported cases are interesting for at least two reasons. First, they show the aggressivity and metastatic potential of MCC. Although the operation in the first patient was performed with wide lateral and deep margins free of tumor, and despite the absence of tumor in the resected neck lymph nodes, the patient developed metastatic disease six months after the initial operation.

The second important point is immunosuppressive therapy, which the first patient had been receiving for two years. Several authors reported the occurrence of MCC in patients receiving immunosuppressive therapy, pointing to this kind of therapy as a probable etiologic factor. According to literature, and in this case, the withdrawal of immunosuppression did not bring any benefit to patient after developing MCC.

Although the second patient had a more advanced disease with regional metastases, he has no evidence of disease 22 months of the treatment. When speculating about the difference between these two patients' outcomes, one can find at least two important points. One point is that the first patient refused recommended adjuvant therapy, and another point is that she must have had a compromised immune system after two years of immunosuppressive therapy, although this therapy was stopped after the diagnosis of MCC.

Upon these two cases one might conclude that surgery alone, albeit radical, gives no satisfying results, although neither adjuvant chemo- or radiotherapy brings much improvement regarding survival according to the literature. Several cases of spontaneous regression contribute to the statement of unpredictable clinical behavior of MCC.

Complete spontaneous regression of MCC was first described in 1986. Since then 10 other cases have been reported. When complete spontaneous regression occurred, it was swift and dramatic with complete regression of skin and lymph node metastases in 1-3 months and no recurrences had occurred[39,40].

There are few reports of nonstandardized treatment for MCC worthy of note. In 3 reported cases intratumoral administration of human tumor necrosis factor was used with success[41,42]. There is one reported case of complete resolution of in-transit metastases from a MCC in response to treatment with isolated hyperthermic limb perfusion with melphalan[43].

According to literature, the predictors of survival are truncal location[6] and tumor stage – nodal status at presentation[6,25,30,31,34]. For patients with stage I disease, the tumor size at presentation is also a predictor of survival[25]. The reported 5-year disease free survival is 30%[15]. The median survival after the initial diagnosis is 45-54 months[3,36]. Three-year overall survival is 57%[36]. Five-year disease-specific survival is 73%-74%[25,36]. Since there is a (remarkable) difference in prognosis between common MCC and immunocompromised MCC patients[16,21] (also observed by our team), this knowledge prompts us to be especially careful in the latter group of patients. Here a radical treatment appears mandatory for MCC and thus early recognition of any skin change in patients receiving immunosuppressive therapy should result in its removal and pathologic verification.

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Sažetak

FACIJALNI KARCINOM MERKELOVIH STANICA: PRIKAZ DVAJU SLUČAJEVA I PREGLED LITERATURE

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Karcinom Merkelovih stanica je agresivan tumor kože koji nastaje iz neuroendokrinih stanica i ima nepredvidivo kliničko ponašanje te loš klinički ishod. Karcinom Merkelovih stanica je relativno rijedak, s nekih 600 objavljenih slučajeva, no čini se da mu incidencija raste zbog sve većeg izlaganja sunčevom zračenju i imunosupresiji. Sve je više izvješća o karcinomu Merkelovih stanica u bolesnika s transplantatima koji primaju imunosupresivne lijekove. Prikazujemo dva slučaja karcinoma Merkelovih stanica na koži lica, jedan od njih u bolesnice koja je primala imunosupresivne lijekove. Prema literaturi, standarna terapija kod karcinoma Merkelovih stanica sastoji se od opsežne kirurške ekscizije primarnog tumora, disekcije vrata zbog opipljivih čvorova i u većini slučajeva naknadne radioterapije. U ovom radu dajemo pregled novijih izvješća o karcinomu Merkelovih stanica u odnosu na dijagnostiku, klinički tijek i mogućnosti liječenja.

Ključne riječi: *Karcinom – Merkelova stanica, etiologija; Karcinom – Merkelova stanica, terapija; Kožne neoplazme, terapija; Presađivanje organa, terapija*