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Review

Merkel Cell Carcinoma: Case Report

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A B S T R A C T

Merkel cell carcinoma (MCC) is a rare, aggressive neuroendocrine carcinoma of the skin. Although it is 40 times less common than malignant melanoma, its mortality is much higher compared to melanoma. From 1986 to 2001 there was rapidly increasing incidence in reported cases of MCC, with a tripling in the rate over this 15-year period. The vast majority of MCC presents on sun-exposed skin. The head and neck area is the most common site of tumor occurrence. We present 70-year old female patient with painless red-colored nodule, size 2x2x2 cm on the dorsal side of mid left forearm. The surgical excision with negative margins was performed, and pathohistological analysis confirmed Merkel cell carcinoma. Sentinel lymph node biopsy was negative. In conclusion, as MCC is a very aggressive rare skin carcinoma with lethal outcome, it should be mandatory to perform biopsies of any suspected skin lesion.

Key words: *carcinoma, merkel cell, photosensitivity, skin neoplasms*

Introduction

Merkel cell carcinoma (MCC) is an aggressive neuroendocrine carcinoma with a propensity for local recurrence, regional lymph node metastasis, and fatal metastatic disease. Toker was the first to describe this tumor in 1972[1]. It is the skin cancer with a mortality of approximately 33% at 3 year, higher than that of melanoma (approximately 15%)[2]. Data from epidemiology show a three-fold increase in MCC from 0.15 to 0.44 per 100 000 annually from the years 1986 to 2001[3]. Several factors likely contribute to this including an aging population, increased aggregate sun exposure and a higher number of immune suppressed individuals. The diagnosis of MCC is rarely made clinically. These lesions are often mistaken for basal cell carcinomas, cysts, squamous cell carcinoma, and cutaneous lymphoma. Furthermore, the advent of the immunohistochemical marker cytokeratin-20 (CK-20) improved recognition of this disease and demonstrating the characteristic »neurosecretory granules within cytoplasmic extensions« [4]. Treatment of MCC is controversial. The surgical excision with negative margins is the first treatment and adjuvant local irradiation now is well established but regional adjuvant (lymph nodes dissection or radiation therapy) remains discussed[5].

We present a 70-year old female patient with skin nodule, size 2x2x2 cm on the dorsal side of mid left forearm. The surgical excision with negative margins was performed, and pathohistological analysis confirmed Merkel cell carcinoma. Sentinel lymph node biopsy was negative.

Case Report

Six months prior to hospital admission a 70-year old female noticed painless red – colored skin change on her left arm. In March 2009 she was referred to a dermatologist due to rapid growth of skin nodule (tumor). At the first visit painless red-colored skin nodule on the dorsal side of mid left forearm, size 2x2x2cm was found (Figure 1a and b). The patient had no history of fever, sweats, weight loss or fatigue. She has diagnosed hypertension and non-insulin-dependent diabetes mellitus for eight years. The patient was admitted to our Department of Dermatovenerology for further examinations. Routine laboratory tests revealed only sideropenic anemia and hyperglycemia. Therefore gastrointestinal endoscopic

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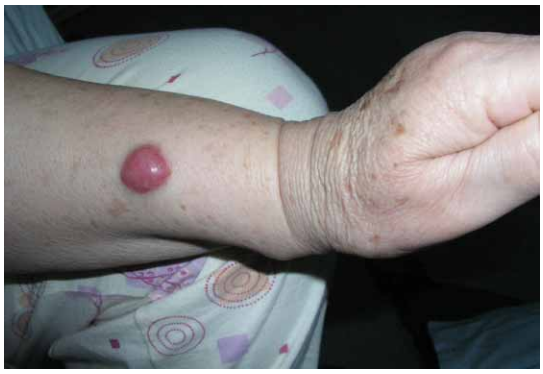
a)



b)

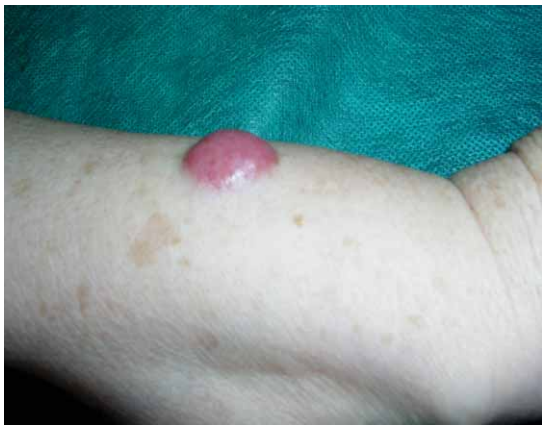


Fig. 1. a) and b) 70-year old female patient with skin nodule on the dorsal side of mid left forearm (Merkell cell carcinoma)

examinations were performed and revealed ventricular erosions while tumor was excluded. In addition, stool tests for occult bleeding were negative, as well as tumor markers. Chest radiograph and abdominal ultrasound findings were normal.

The skin tumor was surgically removed completely and pathohystological diagnosis was Merkel cell carcinoma (MCC). Immunohystochemical staining showed tumor cells positive for neuron-specific enolase (NSE) with perinuclear expression of cytokeratin 20 and neurofilament. Sentinel lymph node biopsy was negative; accordingly disease was localized on primary skin lesion (stage II).

Regular appointments for skin and lymph nodes examinations were performed every 3 months, without recurrence of skin changes, so no additional therapy was initiated.

Discussion

Merkel cell carcinoma (MCC) is a rare, highly aggressive skin cancer with rapidly increasing incidence during last few decades[3]. Similarly, incidence of melanoma is increasing worldwide as well in Croatia, reported by Buljan et al[6]. Mortality of MCC is approximately 33% at 3 years[2], higher compared to melanoma (15%). The tumor is most commonly found in sun exposed areas (head and neck), suggesting ultraviolet radiation as a possible etiologic factor Other risk factors as imunosuppression and viral infection could play role in pathogenesis of MCC as there are reports of MCC cases in kidney transplant patients[7,8]. The most significant features can be summarized in an acronym AEIOU – Asymptomatic, Expanding rapidly, Immune suppression, Older than age 50, and UV-exposed site in person with fair skin[9]. The group of American Joint Committee on Cancer recently proposed a modified 4-tiered system that separates patients with localized disease into stage I (primary tumor dimension less then 2cm) and stage II (primary tumor dimension larger or equal 2cm). Patient with regional or distant metastatic disease are classified as stage III and IV [2].

Our 70-year old female patient presented with growing skin nodule size 2x2x2cm on the left forearm. The skin tumor was surgically removed completely and pathohystological diagnosis was Merkel cell carcinoma (MCC). Immunohystochemical staining showed tumor cells positive for neuron-specific enolase (NSE) with perinuclear expression of cytokeratin 20 and neurofilament. Sentinel lymph node biopsy (SLNB) is generally accepted method in determining status of lymph nodes in patients with malignant melanoma[10], hence SLNB was performed in our patient to obtain staging and prognosis of MCC. SLNB was negative; therefore disease was localized on primary skin lesion (stage II). There is no explicit algorithm for local radiological therapy in MCC stage I and II [11]. We confer with oncologists which recommended no additional therapy for this patient after surgical excision of skin tumor. During the follow up in next 8 months there was no recurrence of tumor; and so we are planning to perform regular visits every 3–6 months.

Conclusion

MCC is a very aggressive rare skin carcinoma with lethal outcome, therefore it should be mandatory to perform biopsies of any suspected skin lesion.

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KARCINOM MERKELOVIH STANICA: PRIKAZ SLUČAJA

S A Ž E T A K

Karcinom Merkelovih stanica (MCC) je rijedak agresivni tumor kože koji nastaje iz neuroendokrinih stanica. Iako je 40 puta rjeđi od malignog melanoma, MCC ima znatno viši mortalitet. U periodu od 1986. do 2001. godine zabilježen je brzi porast incidencije MCC, tako da se u posljednjih 15 godina incidencija utrostručila. Velika većina MCC se javlja na fotoeksponiranim dijelovima kože, naročito na glavi i vratu. Prikazali smo 70-godišnju bolesnicu s bezbolnim crvenkastim nodusom, veličine 2x2x2 cm na dorzalnoj strani srednjeg dijela lijeve podlaktice. Učinjena je kirurška ekscizija tumora u cijelosti (negativni rubovi), te je patohistološkom dijagnozom potvrđen MCC. Odstranjen je i sentinal limfni čvor koji je ispao negativan. U zaključku, budući da je MCC rijedak, vrlo agresivan kožni tumor sa smrtnim ishodom, potrebno je učiniti biopsiju svake sumnjive kožne promjene.