Merkel Cell Carcinoma of Skin in the Thigh: A Case Report on Metastasis to Groin and Abdominal Wall Without Involvement of Viscera

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Abstract: Merkel cell carcinoma (MCC) is an exceptional neoplasm that occurs in the age group of 60-80. It involves the skin with an aggressive behavior, including visceral metastasis, recurrence and fatal outcome. The most common etiologies include exposure to sun, cumulative immuno suppressant, age, virus and genetic alterations. The therapeutic guidelines for MCC are yet to be established, however; either local or regional surgical intervention or adjuvant radiotherapy are the primary therapeutic options, which improve both loco-regional control and disease-free survival. The present study on MCC is found worthwhile to focus in view of the following observations which are contrary to the reports published in the literature like; observance of MCC at the age of 38 years against the general observation at 60-80 years, occurrence at thigh against the common involvement of upper parts of the body, occurrence at a part of body which is not exposed to sun and metastasis to inguinal region and abdominal wall without involvement of viscera. A 38-year-old male presented with a swelling in left thigh. As a first-line therapy excision biopsy revealed MCC. Stage was I-B. He presented 3 months later with swellings in inguinal region and left abdominal wall. FNA of both inguinal and abdominal wall was consistent with MCC. The patient was referred for adjuvant local radiotherapy due to the metastatic nature of disease. Further work on epidemiology of MCC is required to confirm the different nature of characteristics in this particular case.

Key words: Merkel Cell Carcinoma · Thigh · Groin · Abdominal Wall · Surgery · Radiotherapy

INTRODUCTION

Merkel cell carcinoma (MCC) or neuroendocrine carcinoma is a rare skin malignancy first described in 1972 [1] and occurs mostly in the age group of 60-80 years [2]. Merkel’s cells are present in epidermis, dermis, nail and oral mucosa and are thought to be originated from the pleuripotential basal cells of the epidermis. These cells are innervated clusters functioning as neurosensory transmitters in the skin [3]. MCC is a highly malignant neoplasm with an aggressive behavior including involvement of a high frequency of lymph nodes, recurrence, metastases and fatal outcome [4]. The presentations are often with painless erythematous to violaceous nodules on sun exposed skin. The tumors ulcerate and grow rapidly within first few months [5]. The locally invasive cancer frequently metastasizes to lymph nodes, liver, lungs, bone and brain [6]. At least 21% of the cases develop distant metastases [7] and the disease specific 5-year survival rates for metastatic MCC are approximately 11% [8]. The most common predisposing factors of MCC include exposure to ultraviolet radiation, organ transplantation, cumulative immuno suppressant, age, skin type, virus detection and genetic alterations [4, 9-11]. Due to its rarity, the therapeutic guidelines of MCC are not well established and hence, either local or regional surgical intervention and adjuvant radiotherapy or radiotherapy alone are the primary therapeutic options and improves both loco-regional control and disease-free survival. The current literature trends do not support the use of chemotherapy [10, 12-15].

The present study on MCC is found worthwhile to focus in view of the following observations which are contrary to the reports published in the literature. (i) Observance at the age of 38 years against the general observation at 60-80 years (ii) Occurrence at thigh against the common involvement of upper parts of the body (iii)
Occurrence at a part of body which is not exposed to sun and (iv) metastasis to inguinal region and abdominal wall without involvement of viscera.

**Case Report:** A 38-year-old male presented with a swelling in left thigh, close to buttocks. The clinical characteristics of MCC revealed asymptomatic/lack of tenderness. The case history of the patient prompted an extensive investigation and as first-line therapy excision biopsy revealed MCC. Definitive diagnosis of MCC was done using ancillary techniques, including electron microscopy and immunohistochemistry [16]. Stage according to Yiengpruksawan system was I-B as tumor was 4.5 cm with no inguinal lymph node or distant metastasis. Adjuvant radiotherapy was not given. The patient presented 3 months later with swellings in inguinal region and left abdominal wall close to umbilicus. There was no metastasis in chest, abdomen, pelvis and bone. FNA of both inguinal and abdominal wall was consistent with MCC. Wide local excision of inguinal and abdominal swelling with a clear margin of 3.5 cm was done and histopathology confirmed MCC at both sites. The patient was referred for adjuvant local radiotherapy for the metastatic nature of disease.

**DISCUSSION**

Merkel cell carcinoma in this case was observed in a male patient aged 38 years who presented with a swelling in left thigh. Age has been reported as one of the major etiological factors in MCC. Generally, the reported age in these patients is 60-80 years, however; age (38) in this particular case did not match reported advanced group of the afflicted [2, 14]. Furthermore the site of MCC in thigh is averse to the most common position in the upper parts of the body. Although MCC is most commonly found on sun exposed areas, it is reported to occur also on trunk, genitalia and perianal regions, which are not exposed to the sun [16]. The occurrence of primary MCC on thigh and its metastasis to abdominal wall refutes the literature reports on the site of MCC in sun exposed localized parts of the body.

Literature reports suggest that MCC presentations are often with painless erythematous to violaceous nodules on sun exposed skin. The tumors ulcerate and grow rapidly within first few months [5, 17]. However, the clinical characters and history presented in this case showed asymptomatic/lack of tenderness and expanding tumor. Our observation confirms the finding of Heath et al. [5] who define identical clinical features of MCC.

The most common predisposing factors of MCC include exposure to ultraviolet radiation, organ transplantation, cumulative immunosuppression, age, skin type, virus detection and genetic alterations [4, 9-11]. Nevertheless, in this particular case, the different aforesaid etiologies, except immunosuppression are ruled out.

Subsequently, 3 months later, the patient presented with swellings in inguinal region and left abdominal wall close to umbilicus. The time of relapse was close to that observed by Veness and Howle, [14]. The locally invasive cancer frequently metastasizes to lymph nodes, liver, lungs, bone and brain [6]. At least 21% of the cases develop distant metastases [7]. However, in this particular case, the MCC metastasized to groin and abdominal wall without involvement of the viscera. Our observation confirms information in the literature [14] which reported MCC occurrences in the ipsilateral nodes of the axilla or groin in most of the relapses.

It is interesting to note that the present study included total management of the patient by intervention with surgery, adjuvant radiotherapy and/or radiotherapy alone. This is in conformity with reports in the literature. Earlier studies considered surgery as the first-line therapy supplemented with adjuvant radiotherapy [15]. This report is further confirmed by Steinstraesser et al. [18] who recommended surgical excision, pathological confirmation and complete removal of the tumor. The authors further emphasized that an early detection of MCC may not require postoperative radiation therapy. In our present study, at no point, chemotherapy was administered. This is in accordance with the literature trends which do not support the use of chemotherapy [15].

**CONCLUSION**

Our study on present case has refuted the literature reports on age as one of the major factors for infliction with MCC and also exposure to ultraviolet radiation as the possible etiology. The primary site of the tumor in this case is thigh, which is averse to common position in the upper parts of the body. This investigation ascertains that MCC can be simply an expanding tumor, lacking tenderness, instead of erythematous to violaceous nodules observed in some studies. Contrary to the universal observation of MCC protruding the viscera, in this particular case we observed metastasis from thigh to inguinal region and abdominal wall without involving the viscera. Most common predisposing factors (ultraviolet radiation exposure, organ transplantation,
cumulative immunosuppression, age, skin type, virus detection and genetic alterations) of MCC, except immunosuppression, are ruled out in this case. The investigation confirms reports in the literature on management of MCC by surgery and radiotherapy. It reiterates and confirms the futility on use of chemotherapy. Further work on epidemiology of MCC is required to confirm the different nature of characteristics in this particular case.

REFERENCES