

CASE REPORT

Merkel cell carcinoma and multiple basal cell carcinoma in an African albino woman with HIV infection

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A 25-year-old HIV-infected African albino woman developed an aggressive Merkel cell carcinoma on her face and at least 10 basal cell carcinomas, mainly on sun-exposed parts of her body. HIV infection, immune deficiency and sun exposure are known risk factors for the development of Merkel cell carcinoma. Chemotherapy and radiotherapy were only temporarily successful. She died shortly after surgery was performed to remove the tumour.

Keywords: Africa, albinism, basal cell carcinoma, HIV infection, Merkel cell carcinoma

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Introduction

Merkel cell carcinoma (MCC) is a rare skin cancer of neuroendocrine origin which affects mainly elderly Caucasians, and which is probably related to sun exposure and immunosuppression status [1]. It is a highly aggressive and lethal tumour. MCC is mainly found on the sun-exposed areas of the head, neck, arms and legs [2]. The tumour is located in the dermis and can spread into the subcutaneous fat and, in rare cases, also into the muscle tissue. Clinically these tumours are usually painless, firm, raised, nodular masses that are red, pink or occasionally blue in colour. The overlying skin is usually intact; however, in advanced lesions the skin may be ulcerated. MCC occurs more commonly in organ transplant recipients and in patients with B-cell lymphoid neoplasm than in the general population [3,4].

Recently it was suggested that MCC also occurs more often in persons with HIV infection [5]. In a search of the AIDS and cancer registers of the USA (1978–1996), ten MCC cases were identified as occurring in both registers [5]. In four of these cases, the MCC was diagnosed before the patient developed AIDS. In the remaining six cases, the MCC was diagnosed in persons with AIDS, corresponding to a relative risk of 13.4 compared with the general population [5].

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MCC is especially uncommon in Black people [1]. We describe an HIV-infected African albino woman with MCC and multiple basal cell carcinomas.

Case report

Over a period of 4 months, a 25-year-old HIV-infected African albino woman, a refugee from Angola, developed a rapidly growing nodular swelling at the level of her right cheek, together with enlarged cervical and submandibular lymph nodes (Fig. 1). On physical examination, at least 10 hyperkeratotic skin lesions were observed on the upper part of her chest, her shoulders and her face (sun-exposed parts of her body). The large swelling on her face was painful but she had no other complaints. Her CD4 lymphocyte count was 332 cells/μL and her viral load was 187 000 HIV-1 RNA copies/mL plasma. A biopsy of the tumour on her face confirmed the diagnosis of MCC and biopsies of two different hyperkeratotic skin lesions revealed the presence of basal cell carcinoma.

In February 2002, she received carboplatin-etoposide treatment. Because of disease progression in March 2002, she was switched to doxorubicin-ifosphamide-cisplatin, once every 3 weeks. After four cycles of chemotherapy a partial response was obtained. Then, in June 2002, consolidative radiotherapy was started. Because of local progression during the radiotherapy, gemcitabine once weekly as a radiosensitizer was added to the radiotherapy. For the HIV infection she initially received the following highly active antiretroviral treatment (HAART): abacavir 300 mg twice a day (bid), nevirapine 200 mg bid and stavudine 30 mg bid. During chemotherapy this treatment

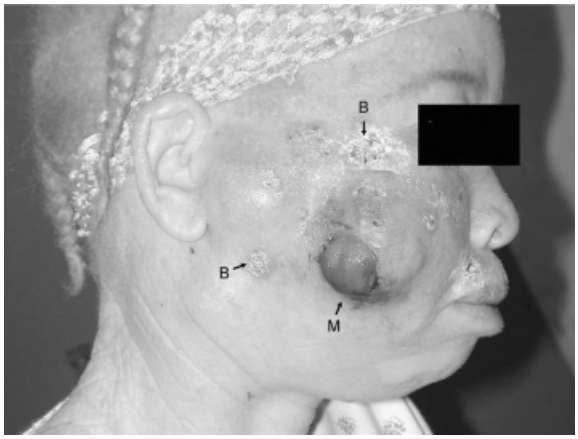

Fig. 1 Merkel cell carcinoma (M) and basal cell carcinoma (B) in the African albino woman.

was stopped because of nausea. There was further local progression and palliative care was started. Finally, in another hospital, surgery was performed to remove the tumour, but in January 2003, 1 month after this intervention, she died.

Discussion

Several cases of MCC in persons with HIV infection have been described [6–10], but as far as we know this is the first African person with HIV infection to have been reported to develop MCC. A review of the literature revealed only one case report of MCC in an African person [10], a 20-year-old Senegalese woman who developed nodular lesions on her foot which were diagnosed as MCC. In her case the tumour was also very aggressive and rapidly metastasized the lymph nodes and the lungs. It was not reported whether this woman had a concomitant HIV infection. Our patient probably developed MCC as a consequence of immune deficiency caused by HIV and because she was an albino living in Africa and frequently exposed to the sun. Albino individuals are known to be at high risk for skin cancers, including basal cell carcinomas, when exposed to ultraviolet radiation (UVR) [11,12]. MCCs are often associated with other neoplasms such as squamous cell carcinoma, Bowen's disease, sweat gland tumours and basal cell carcinomas [13]. In the USA, the increased incidence of MCC observed among persons with AIDS, compared with the general population, may be related to the immune deficiency of HIV-infected individuals but also to greater exposure to UVR in these individuals. Homosexual men in particular have been reported to have higher exposure to recreational UVR than HIV-seronegative controls without risk factors for HIV infection [14]. Of the 10 individuals diagnosed with HIV infection and MCC between 1978 and 1996 in the USA, at least five but possibly up to nine were homosexual/bisexual men [5]. Not all of these patients had severe immune deficiency, because in four of them the MCC developed before the AIDS diagnosis. Our case report supports the theory that, in HIV-infected individuals, not only immune deficiency but also exposure to UVR is a risk factor for developing MCC.

As in other reported cases, the MCC in our patient was particularly aggressive. Because of the rare occurrence of MCC, randomized clinical trials evaluating different treatment approaches have never been carried out. Therefore, there is still no standardized treatment for this tumour. If the tumour is still small and if there is no lymph node involvement, surgical removal followed by radiation therapy, with or without chemotherapy, is indicated [15]. Because of the highly invasive nature of the tumour, wide local excision with a 2–3 cm margin of normal tissue is recommended [16]. When there are distant metastases, chemotherapy will only be palliative and the survival period will be short with only a few brief remissions. This was the case in our patient.

Our patient presented with at least 10 basal cell carcinomas. An increase in this type of skin cancer has also been observed in persons with HIV infection [17]. In individuals with severe immune deficiency, the basal cell carcinomas seem to grow more rapidly and be more invasive than in other groups of patients [18].

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