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Case Presentation
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Merkel Cell Carcinoma of the Eyelids – One Centers Experience

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Abstract

We report three cases of merkel cell carcinoma (MCC) of the eyelids treated at East Tallinn Central Hospital Eye Clinic. The records of three patients with histologically proven MCC were analyzed retrospectively. Recorded data included demographic features, type of treatment and outcome. They were seen and managed by the third author (AK) from 2002-2016. None of the patients demonstrated clinical evidence of lymph node involvement or metastasis at the time of initial presentation. Each case was staged according to initial clinical presentation using AJCC TNM staging for MCC, 7th edition. All patients were treated with wide surgical excision. One patient received additional radiotherapy because of tumor free margins of only 3 mm. All the patients are under observation without recurrence since wide excision of the tumor was done.

Keywords: Merkel cell carcinomal; Eyelid tumor

Introduction

Merkel cell carcinoma of the eyelids is a rare, highly malignant aggressive neuroendocrine tumor of the skin with the potential for regional and distant metastasis [1-3]. Merkel cells were believed to arise from epidermal tissue with neuroendocrine differentiation [1-3], but recent literature contends that these tumors often exhibit non-epithelial cell and sarcomatous features, which suggests a stem cell origin [3]. These neoplasms typically develop in elderly individuals on sun exposed areas in the head, neck, and upper extremities [2,4]. There is also a strong association of MCC with history of infection with polyoma virus and immunosuppression [3,5]. It has been reported to have a high recurrence rate after excision [1]. Of all MCC cases, 5% to 10% occur on the eyelids [6]. Compared with MCCs occurring in other locations, MCCs of the eyelid appear to be associated with a better prognosis, which may be related to earlier detection [6].

Clinically, the tumor appears as a purple-red "violaceous" vascularized painless cutaneous nodule with occasional ulceration [1,7,8]. Superficial telangiectasis may be present [2]. Eyelid MCCs have a strong predilection for the upper eyelid, often involving the lateral portion of the eyelid and the eyelashes at the anterior eyelid margin [2,8]. MCC in the eyelid area is commonly misdiagnosed initially as cysts, chalazia, or basal cell carcinomas probably because of its low incidence [1-3]. Histologically, MCC is often misdiagnosed as lymphoma, melanoma, or metastatic small cell carcinoma of the lung (SCCL) [9].

Case Report No. 1

A 62-year-old woman had an enlarging tumor on her left upper eyelid for 6 weeks. Greatest dimension of the tumor was 25 mm and the tumor was classified as T2N0M0 tumor and underwent radical resection with 5 mm tumor free margins. The reconstruction of the defect was done with a pedunculated flap skin graft taken from the ipsilateral side lower eyelid. Histologically MCC was confirmed. She has been 14 years under observation without recurrence (Figures 1-3).

Case Report No. 2

A 71-year-old woman had an enlarging reddish tumor on the left lower eyelid for 6 weeks. Greatest dimension of the tumor was 18 mm and the tumor was classified as T1N0M0 tumor and she underwent tumor excision with lower lid transpositional flap from the temporal side. Histologically MCC was confirmed. She has been tumor free for 3 years (Figures 4-6).

Case Report No. 3

A 82-years-old woman had a small reddish tumor on the left lower eyelid for 2 months,

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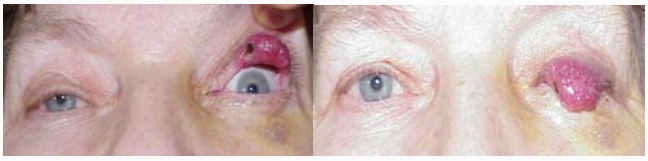

Figure 1: Patient no. 1with an enlarging tumor on her left upper eyelid (A & B).

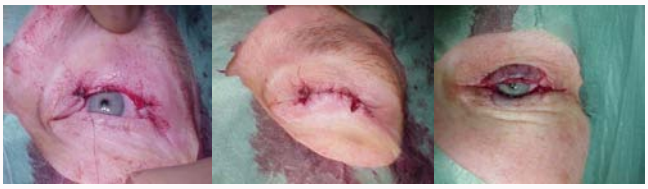

Figure 2: A & B) Radical resection of the tumor with 5 mm tumor free margins. The reconstruction of the defect was done with a pedunculated skin graft flap taken from the ipsilateral side lower eyelid, using modified Cutler-Beard technique. C) 4 weeks after the first operation second operation was carried out for the eyelid reconstruction.

Figure 3: Postoperative result 8 weeks (A) and 14 years (B) after the operation.

Figure 4: Patient no. 2 with a reddish tumor on the left lower eyelid.

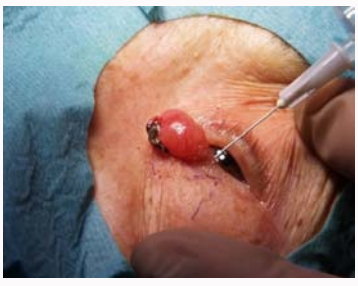

Figure 5: Tumor was excised with 5 mm tumor free margins. Reconstruction was carried out with lower lid transpositional pedunculated flap from the temporal side.

which was excised by a general surgeon. No histologic examination was performed because the tumor was thought to be benign. After tumor´s removal it grew back after 3 weeks. Thereafter an ophthalmo

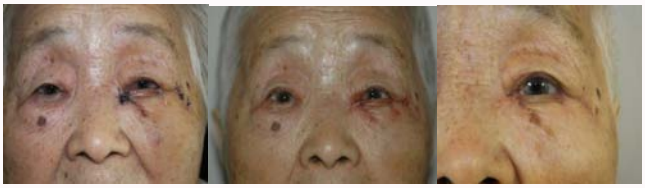

Figure 6: Postoperative result 10 days (A), 20 days (B) and 2 months (C) after the operation.

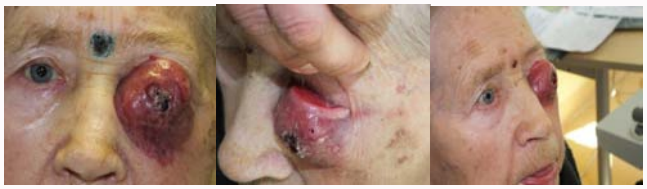

Figure 7: Preoperative pictures of patient no. 3 with rapid growth of the tumor after first excision of a small tumor of the left lower eyelid (A, B & C).

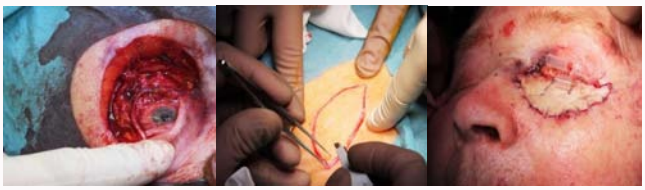

Figure 8: Wide excision of the tumor (A). Reconstruction was done with a free skin transplant from the medial brachium (B, C).

Figure 9: Postoperative result 7 days (A), 14 days (B) and 9 months (C) after the operation.

surgeon was consulted, who had a suspicion of MCC. A thorough examination was then also performed by an oncologist without any signs of general spread of a tumor. Greatest dimension of the tumor was 55 mm and the tumor was classified as T3N0M0 tumor. After wide surgical excision of the tumor, reconstruction was done with a free skin transplant from the medial brachium. Histologically MCC was confirmed. Due to tumor free margins of only 3 mm, radiotherapy was added after surgical excision. The patients follow up time has been 9 months without a recurrence (Figures 7-9).

Discussion

MCC is a rare malignant disease, but the clinical features of the tumor help to make the diagnosis. Due to the rapid growth and aggressiveness of the tumor a multidisciplinary approach is needed. In the literature histologically proven complete spontaneous regression after non-radical excision of the tumor has been described the immunologic phenomenon of spontaneous regression points out the importance of the immune system in this disease [10].

All our patients were female, the lesion was on the left side and they complained of a fast growing tumor with no pain or itching. The tumor was bleeding when manipulated and patients were also

consulted by an oncologist to exclude systemic spread.

A 5 mm margin with histologic confirmation of tumor-free margins would seem reasonable for local control of eyelid merkel cell carcinoma and is consistent with management of other types of aggressive malignant eyelid lesions [1,3]. The margin of excision should account for all cutaneous margins, as well as the deep and subcutaneous margins of the tumor [2]. Frozen-section monitoring of this 5 mm margin can be performed intra-operatively [2] and Mohs micrographic surgery has been shown to be an appropriate approach ensuring margins free of disease [3,4,11]. There are currently no universally accepted protocols for the treatment of merkel cell carcinoma, in part because few institutions have collected enough patients to allow randomized trials of treatment modalities [1,3,11]. Wide surgical excision of eyelid MCC and eyelid reconstruction currently seems to be the standard of care as demonstrated by review of the literature, more aggressive prophylactic therapy may be indicated in selected cases [1]. Some studies suggest a need to take a sentinel lymph node biopsy in patients with MCC of eyelid and periocular region [3]. Some authors suggest that the use of radiotherapy should be reserved for tumor recurrences or regional lymph node spread [2], but others recommend adjuvant prophylactic irradiation of the tissues between the tumor and first regional lymphatic nodes (50 Gy to 60 Gy in 20-25 fractions — except for patients who have small tumors and no evidence of lymphatic or lymph node involvement), because the MCC is a neoplasm highly sensitive to ionizing radiation therapy [12]. Postoperative radiotherapy for tumor site and regional lymph nodes has significant importance for local control of the disease, but no impact on the incidence of metastatic disease and survival rate [12]. The most common sites of distant spread are the liver, lungs, brain, and bones [2,8,13]. MCC of the eyelid has been reported to have an overall metastatic rate ranging from 10% to 30%, regional lymph node recurrence rate of 20%, and distant metastasis rate of 5% [3]. Systemic metastasis and extensive regional nodal metastasis have been treated with a variety of chemotherapeutic regimens, but the general prognosis for patients with metastatic merkel cell carcinoma is poor [2,3]. MCC has demonstrated sensitivity to drugs such as cisplatin, cyclophosphamide, doxorubicin, vincristine, and 5-fluorouracil [3]. Adjuvant chemotherapy has not been proven to diminish the rates of recurrence nor improve survival [3]. The poor prognosis for patients with disseminated MCC emphasizes the need for early diagnosis and appropriate treatment with wide surgical excision [2]. Lifetime follow-up for patients treated for eyelid MCC is recommended [2].

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