

Merkel Cell Carcinoma of the Upper Eyelid: When Reconstruction Becomes a Challenge

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Merkel cell carcinoma (MCC) is a rare and aggressive tumor arising from Merkel cells located in the basal layer of the epidermis and in the hair follicle. MCC presents primarily in Caucasians (98.3%) with a median age of 69 years. Ultraviolet radiation exposure and immunosuppression appear to play a role in the development of this cancer [1]. MCCs display an aggressive clinical course with a high rate of local recurrence (26%–60%), regional lymph node invasion (45%–91%), and metastasis to the lung, liver, brain, bone, and skin (18%–52%). Eyelid MCCs account for 5%–10% of all MCCs [2]. Although clinical management is often discussed, few cases involving the eyelid location have been described. For anatomical reasons and due to the rarity of the tumor, standard site-specific treatment regimens have not been defined [3]. We present a case of Merkel cell carcinoma of the eyelid that was treated with the aim of achieving surgical eradication of the tumor, as well as satisfactory aesthetic and functional outcomes.

A 75-year-old Caucasian female was referred to our clinic 1 month after presentation of a rapidly growing lesion of the right upper eyelid. Clinical examination showed a solitary, asymptomatic, 1 cm diameter, pink-red vascularized cutaneous nodule of the middle

third of the right upper eyelid (Fig. 1). Histological examination of an incision biopsy established the diagnosis of MCC. Clinically, there was no evidence of lymph node involvement in the head, neck, or axillae. The preoperative staging of MCC included a whole-body computed tomography (CT) spiral scan with contrast and positron emission tomography (^{18}F -fluorodeoxyglucose positron emission tomography/CT). Screening did not reveal any metastases, classifying the patient in IB stage (T1-cN0-M0). Under general anesthesia, we performed a wide excision of the whole upper right eyelid containing the lesion with 1 cm free margins and preserving the upper part of the lid levator muscle. Intraoperative sections showed no tumor presence in the edges. A full thickness fasciocutaneous Fricke flap from the temporal region was raised, and passed subcutaneously, after de-epithelialisation, to reconstruct the upper lid (Fig. 2). The pedicle, based on peripheral branches of the superficial temporal artery, had previously been identified by ultrasonography about 1 cm from the lateral orbit rim. In order to preserve palpebral functionality, the internal lining (i.e., fascia) of the flap was anchored to the levator muscle. Conjunctival reconstruction using a mucosal graft taken from the lower lip was also necessary for the internal lining. Histologic examination showed a MCC involving the skin and the conjunctiva of the upper lid positive to CK-20 and synaptophysin, with complete excision. The patient underwent adjuvant radiotherapy. At the 1 year follow-up, the patient was free from disease and continued to demonstrate a good functional and cosmetic outcome (Figs. 3, 4).

MCC is a rare skin cancer with a mortality rate higher than melanoma. In its early stage, the neoplasm does not demonstrate specific clinic characteristics. Given the rarity of the tumor and the possibility of misdiagnosis, a high index of suspicion is necessary, especially in immunosuppressed patients. The



Fig. 1. Preoperative planning of a 1 cm nodular merkel cell carcinoma excision and reconstruction with a Fricke flap. The vascular pedicle is marked in red after hand-held Doppler scanning.

**Fig. 2.**

An intraoperative image showing the tunneled Fricke flap inset after merkel cell carcinoma excision and an oral mucosal graft positioned with direct closure of the donor site along the natural wrinkle.

**Fig. 3.**

Six-month postoperative result after complete functional recovery (eyes wide open).

**Fig. 4.**

Six-month postoperative result after complete functional recovery (eyes closed).

diagnosis is essentially histological. The staging of the tumor necessitates a whole-body CT spiral scan and PET. In stage I and II, wide local excision of the primary lesion with 1 to 2 cm margins investing fascia remains the gold standard. Mohs' micrographic surgery or modified Mohs' surgery can be considered in critical tissue sparing cases such as for facial MCC. Of course, in the lids, excision needs to involve the full thickness of the lid itself. The necessity of a sentinel lymph node is controversial, but it can be helpful in staging and for prognosis. The use of local adjuvant radiation therapy (RT) decreases the risk of recurrences. The plasmatic dosage of chromogranin A and NSE can be useful in post-treatment follow-up. Chemotherapy treatment must be considered in cases of advanced disease. A number of techniques have been described for eyelid reconstruction after a full thickness excision. A Tenzel flap (or semicircular rotation flap) is useful for covering defects of 40%–60% of the eyelid, while in our case we performed a total upper lid excision [4]. The Cutler-Beard, or bridge flap, is the classic eyelid sharing procedure, but it requires prolonged occlusion of the eye and a two-stage procedure (division of the flap traditionally occurs in 6 weeks), and it can produce persistent lower eyelid instability and upper lid entropion. The Mustardé lid flap utilizes a two-stage transposition of the lower lid margin including the lashes. In this case also the flap division occurs in 6 weeks. Further disadvantages include the potential irritation of the

cornea by the pedicle, the need for full-thickness lower eyelid reconstruction, and a poor cosmetic outcome of the lower lid. The Tripiier flap has also been reported to be used in this situation, but it can result in entropion due to the lack of rigid support to replace the tarsus. The forehead flap is generally one of last choice, as it is a very thick flap that requires a two-stage procedure (usually 2 weeks are required for division of the flap). This flap can be combined with various other procedures to repair full-thickness defects of the eyelids [5].

The temporal forehead flap described by Fricke is a transposition flap used for full-thickness eyelid defects. This flap allows lower lid sparing, avoiding prolonged eye occlusion, with a single-step surgical procedure. The vascular pedicle is reliable, and fast and easy to raise and harvest. Usually the flap donor site scar can be easily hidden in the wrinkles or in the brow margin; besides, one step reconstruction of the lashes by using eyebrows can be performed. For these reasons, our choice has ensured long-term eye protection, good functionality, and an aesthetic shape.

Due to the rarity of this tumor, there is no consensus on its optimal treatment, especially with regard to palpebral localization. We conclude that an early and wide local excision of the primary tumor either alone or with RT is actually the more appropriate treatment for stage IA and IB MCC.

Composite full-thickness reconstruction is always required in MCC. For this reason, the physician who

approaches such a case should be confident in performing this kind of flap.

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Cutaneous T-Cell Lymphoma with Nasal Necrosis

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Mycosis fungoides is a rare T-cell cutaneous lymphoma that poses a unique diagnostic challenge given its heterogeneous presentation. The refractory case presented here highlights the need for advances in early diagnostic technologies.

A 73-year-old diabetic patient presented with erythema and necrosis of the nose. He initially had a 7 mm squamous cell carcinoma on the nose which

was excised with Mohs surgery. The wound rapidly progressed to full thickness necrosis of the left nasal ala without pain or fever. During subsequent reconstruction with a nasolabial flap and cartilage graft, biopsies of the nasal margin showed severe inflammation without malignancy. The reconstructed area failed to heal and serial cultures grew multiple species of bacteria which were treated with several courses of antibiotics. Erythema of the nose and face continued to progress over 7 months and new lesions appeared, including skin necrosis at the site of previous left ala flap, right dorsum of nose and right cheek (Fig. 1). Computed tomography (CT) scan showed no involvement of sinuses or facial bones. Repeat biopsy showed acute and chronic inflammation with no malignancy and no organisms. A few weeks later a new erythematous plaque developed on the back (Fig. 2) and biopsy showed lymphocytic infiltrate with a monoclonal T-cell population (Figs. 3, 4). Repeat assay of previous nasal biopsy showed the same T-cell receptor gene rearrangement leading to the diagnosis of cutaneous T-cell lymphoma, consistent with mycosis fungoides (MF). The patient was staged with positron emission tomography-CT scan showing no visceral disease despite developing new lesions in his groin in addition to new lesions on his back and face. He was treated with a combination of systemic chemotherapy, radiation and phototherapy. At 16 month follow up he has persistent skin lesions, with progression of tumor burden.

This case highlights the diagnostic difficulty seen

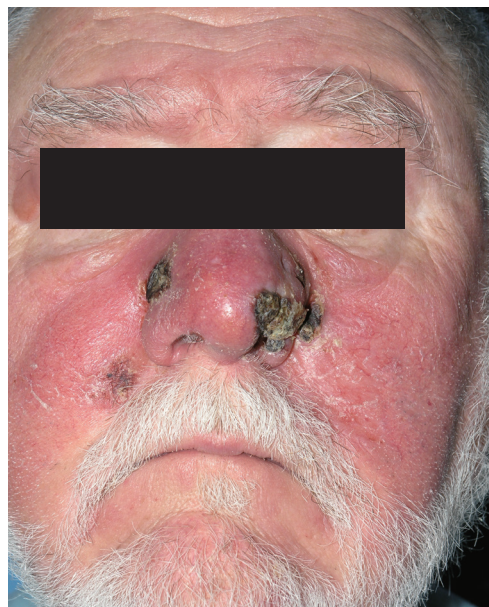


Fig. 1. Facial involvement of mycosis fungoides with significant nasal necrosis at site of previous nasolabial flap.