

Merkel Cell Carcinoma on the Upper Lip of a 100-year-old Woman

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Abstract

Merkel cell carcinoma (MCC) is a rare skin tumor that usually occurs on the head, neck, or extremities of elderly patients; it has a high incidence of local recurrence, regional lymph node metastasis, and subsequent distant metastasis. We report a MCC that developed rapidly on the left corner of the upper lip of a 100-year-old woman. An incisional skin biopsy was performed to confirm MCC. Computed tomography showed no metastasis. The tumor was widely excised with a margin of 1 cm. Immediate reconstruction with a reverse Estlander flap from the lower lip was performed under general anesthesia. Additional surgery was also performed under general anesthesia 2 weeks later to widen the patient's lips. The surgical results were satisfactory. The patient died of senile deterioration a year after hospitalization for long-term medical treatment, without any recurrence or metastasis of MCC. Despite the patient's age, we considered it necessary to resect the tumor widely because of its rapid growth. The tumor margin was 1 cm. No radiotherapy was performed, but we believe that surgery alone was effective in allowing this patient to live an additional year without recurrence or metastasis. To the best of our knowledge, this patient is the oldest person with MCC yet described.

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Key word: Merkel cell carcinoma elderly

Introduction

We report a Merkel cell carcinoma (MCC) that developed rapidly on the left corner of the upper lip of a 100-year-old woman; it was successfully treated with surgery alone. To the best of our knowledge, this patient is the oldest person with MCC yet described.

Case Report

A 100-year-old woman presented with a tumor that had grown rapidly on the left upper lip. Physical examination showed a diffuse, raised red tumor (**Fig. 1**), which was difficult to diagnose macroscopically. MCC was diagnosed on the basis of an incisional skin biopsy. Computed tomography showed no metastasis. The patient's medical history

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included chronic heart failure and chronic sinusitis, but these conditions did not preclude general anesthesia.

During the first operation, the mass was widely excised with a margin of 1 cm (**Fig. 2a, 2b**). Immediate reconstruction was performed with a reverse Estlander flap from the lower lip (**Fig. 2c**). Additional surgery was also performed under



Fig. 1 Tumor at the patient's first visit
A diffuse, raised red tumor can be seen on the upper lip.

general anesthesia 2 weeks later to widen the patient's lips so that she could wear dentures (**Fig. 3**). The results were cosmetically satisfactory, and, because there were no functional disabilities, such as trismus, the patient was able to eat and drink (**Fig. 4**).

On immunohistochemical examination of the lesions, the tumor cells expressed immunoreactivity for cytokeratin 20 and showed a characteristic paranuclear dot-like pattern, confirming our diagnosis of MCC. All surgical margins were negative (**Fig. 5**).

Plans had been made to transfer the patient to another institution for postoperative radiotherapy, but pseudomembranous enteritis developed after the second surgery, making transfer difficult. Because the results were both clinically and cosmetically satisfactory, the patient and her family finally decided against transfer for additional treatment. The patient was later moved to a long-term care institution and died of senile deterioration a year

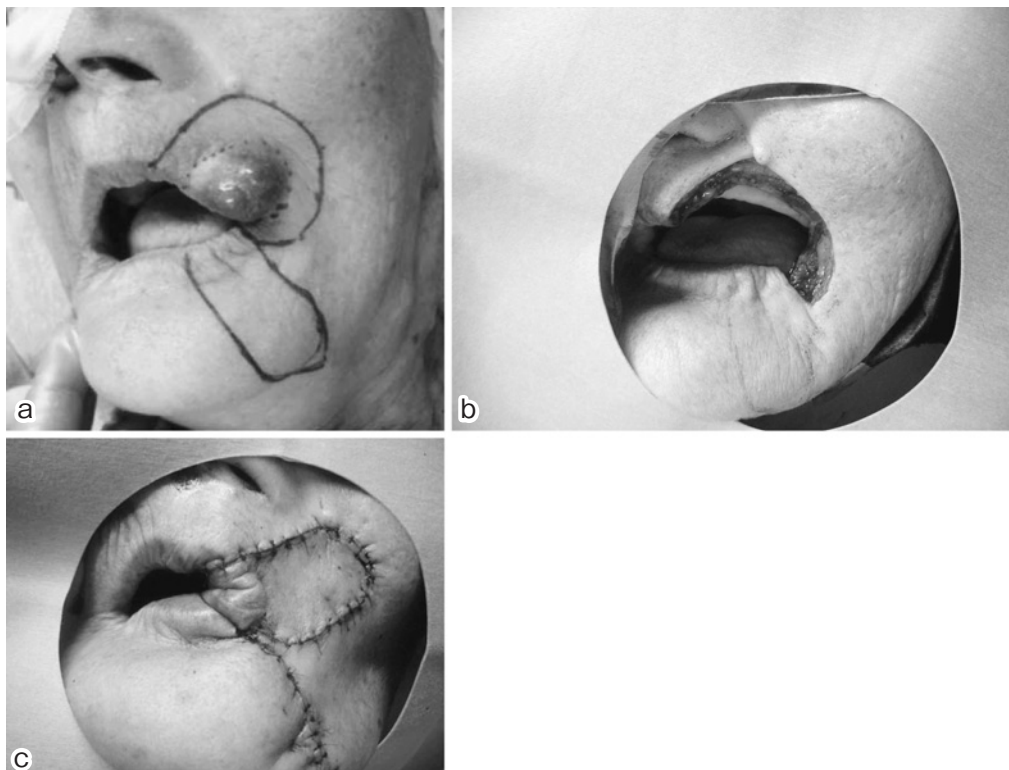


Fig. 2 Intraoperative view during the first surgery
The tumor grew rapidly over a period of 3 weeks.
a: Design of a reverse-Estlander flap
b: After tumor excision
c: After flap transposition

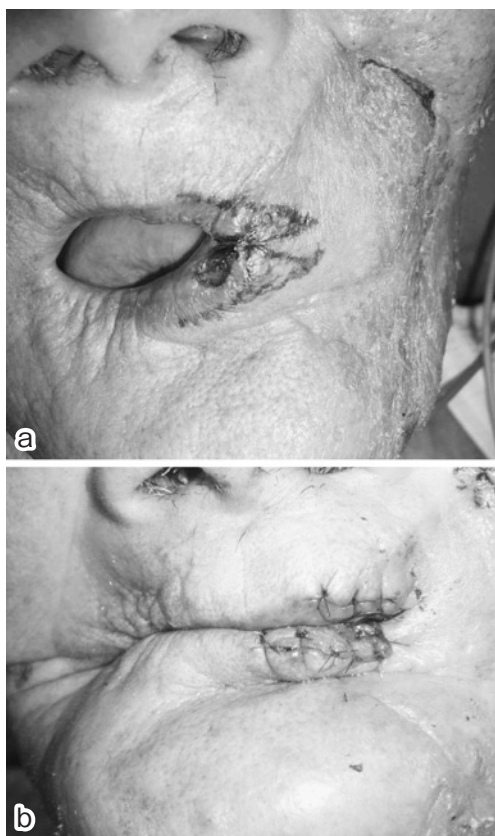


Fig. 3 Intraoperative view during the second surgery
a: Design of chalinoplasty
b: A triangular flap was inserted into the oral cavity, and the vermillion borders of the upper and lower lips were advanced.

later without any recurrence or metastasis of MCC.

Discussion

MCC is a rare skin tumor that usually occurs on the head, neck, or extremities of elderly patients; it has a high incidence of local recurrence, regional lymph node metastasis, and subsequent distant metastasis. The average survival time in Japan for patients with MCC is 2.7 years; so, despite the patient's age in the present case, we believed that the tumor needed to be widely resected because of its rapid growth. Along with these concerns, the patient and her family also desired that she live out her life with minimal facial deformation. Our goal, therefore, was to prevent recurrence within the normal survival period while minimizing facial deformation.

There is no generally accepted staging system for skin cancer. Some use the American Joint



Fig. 4 8-month postoperative view
Functionally and cosmetically satisfactory results were obtained without any recurrence.
a: closed position b: open position

Committee on Cancer Staging System for skin cancer or a modified version of it proposed by Yiengpruksawan et al¹. Wide excisions with margins of 2 to 3 cm are recommended, but these are difficult to achieve in the head and neck²; this was our main concern for this patient. Margins of 3 cm may be ideal for MCC. However, reconstruction of the buccal and nasal regions would also be needed along with reconstruction of the lip, which requires long operations, such the raising and transfer of free flaps. Controversy still exists over the optimal margins of excision, because no controlled trials

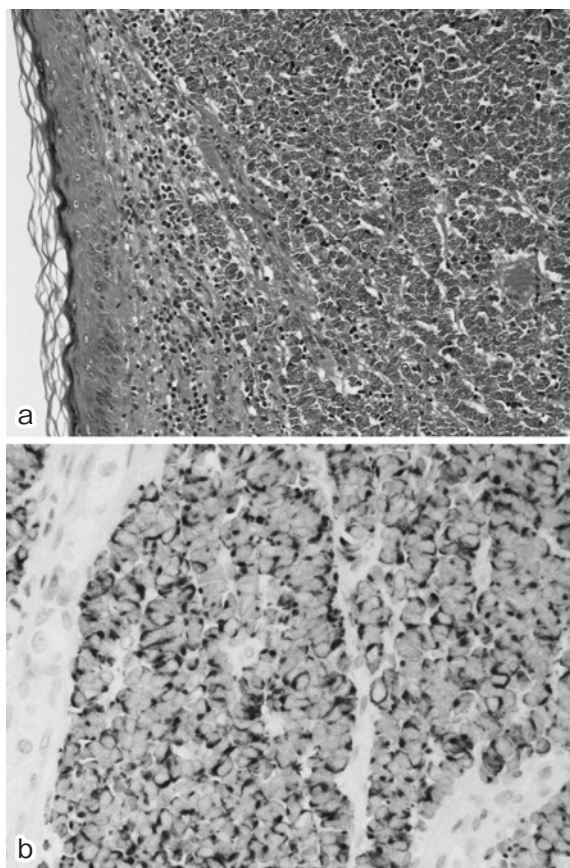


Fig. 5 Histopathological findings
a: Hematoxylin-eosin stain ($\times 100$)
 Small round basophilic cell was seen.
b: Cytokeratin 20 stain ($\times 400$)
 Distinct perinuclear dot-like positivity was seen.

have compared different margins with outcomes^{3,4}. Now that wide-field adjuvant radiotherapy is available, wide excision margins do not necessarily come at the expense of function and form, assuming a negative microscopic margin is obtained⁴. First-line treatment in the case of a localized disease need not involve chemotherapy¹. Considering this information and the patient's age, we planned a resection with a

margin of 1 cm with the aim of acquiring satisfactory results simply by reconstructing the lip and preventing relapse of the tumor with additional radiotherapy.

Immunohistochemical staining was essential to confirm the diagnosis in this patient¹. MCC is believed to be derived from neuroectoderm and frequently expresses neuroendocrine-specific markers, particularly cytokeratin 20 (as in this case), often as paranuclear dots⁵.

Although the planned additional radiotherapy could not be performed because of unexpected complications, the surgical margin of 1 cm was negative, and there was no recurrence of the tumor. We believe, therefore, that surgery alone was clinically and cosmetically effective in allowing this patient to live an additional year without recurrence or metastasis.

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