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CASE REPORT

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Rare and aggressive tumor in an unexpected patient

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ABSTRACT

A 56-year-old female patient presented with complaints of medial swelling in her left upper eyelid, growing rapidly for 3 months. She stated that she had received local antibiotic therapy and steroid therapy in two different centers with the diagnosis of chalazion, but with no benefit. For differential diagnosis, we planned a surgical excision of the tumor and chose the Cutler-Beard technique. The pathological and immunohistochemical results of the mass were reported as Merkel cell carcinoma without lymphovascular or perineural invasion. For tumors with high risk of distant metastasis such as MCC, early diagnosis, choosing the appropriate surgical technique, and adjuvant treatments have a positive effect on quality of life and survival.

KEY WORDS: Merkel cell carcinoma, Cutler-Beard technique, eyelid reconstruction.

INTRODUCTION

Merkel cell carcinoma (MCC) is an aggressive skin malignancy which originates from Merkel cells, the mechanoreceptors of the skin, with a mortality rate of over 40%. This skin cancer is more common among the elderly, particularly those who are exposed to high levels of UV radiation. Polyomavirus and immunosuppression have crucial roles in the pathogenesis of the tumor [1-4].

In cases with suspicion of malignancy, surgical technique is important for the success of functional therapy, aesthetic treatments, and other subsequent treatments. For periocular malignant tumors, a clean surgical margin of 5 mm is recommended for primary tumor control [5, 6]. Many studies have shown adjuvant radiotherapy to increase survival, particularly in large tumors, even in cases of surgical margin negativity. This research was performed in line with the principles of the Declaration of Helsinki. Informed consent to publish the photographs was signed by the patient.

CASE REPORT

A 56-year-old female patient presented with complaints of medial swelling in her left upper eyelid, growing rapidly for 3 months. She stated that she had received local antibiotic therapy and steroid therapy in two different centers with the diagnosis of chalazion, but with no benefit. The patient had no known history of comorbidity.

On examination, best-corrected visual acuity was 20/20 in both eyes with a normal intraocular pressure. She had a red-purple ulcerated mass with telangiectatic vessels, 11 × 16 mm in size on her medial left upper eyelid (Figure 1). Examination of other anterior and posterior segments was unremarkable in both eyes so additional imaging methods such as MRI and USG were not needed considering that the tumor was located only in the eyelid. For differential diagnosis, we planned an excisional biopsy considering pilomatrixoma, sebaceous gland carcinoma, and other malignancies.

We marked a 5 mm clear surgical excision margin around the tumor. After local anesthesia, tumor excision was completed using Stevens scissors and a 15-gauge scalpel. After the surgical excision of the tumor, we chose the Cutler-Beard technique due to the presence of more than a 50% defect in the upper eyelid and the intact levator aponeurosis (Figure 1).

A Cutler-Beard flap was prepared by a horizontal incision 2 mm below the lower eyelid in accordance with the defect size. The flap was advanced under the eyelash side and to the defect area on the upper lid. The anterior and posterior lamellar of the flap were separated and sutured to the levator and orbicularis muscles to cover the defect area (Figure 1).

The pathological and immunohistochemical results of the mass were reported as Merkel cell carcinoma sized 2.1 × 2.1 cm without lymphovascular or perineural invasion (Figure 2). The distance from the tumor to the nearest clean surgical margin was 5 mm.

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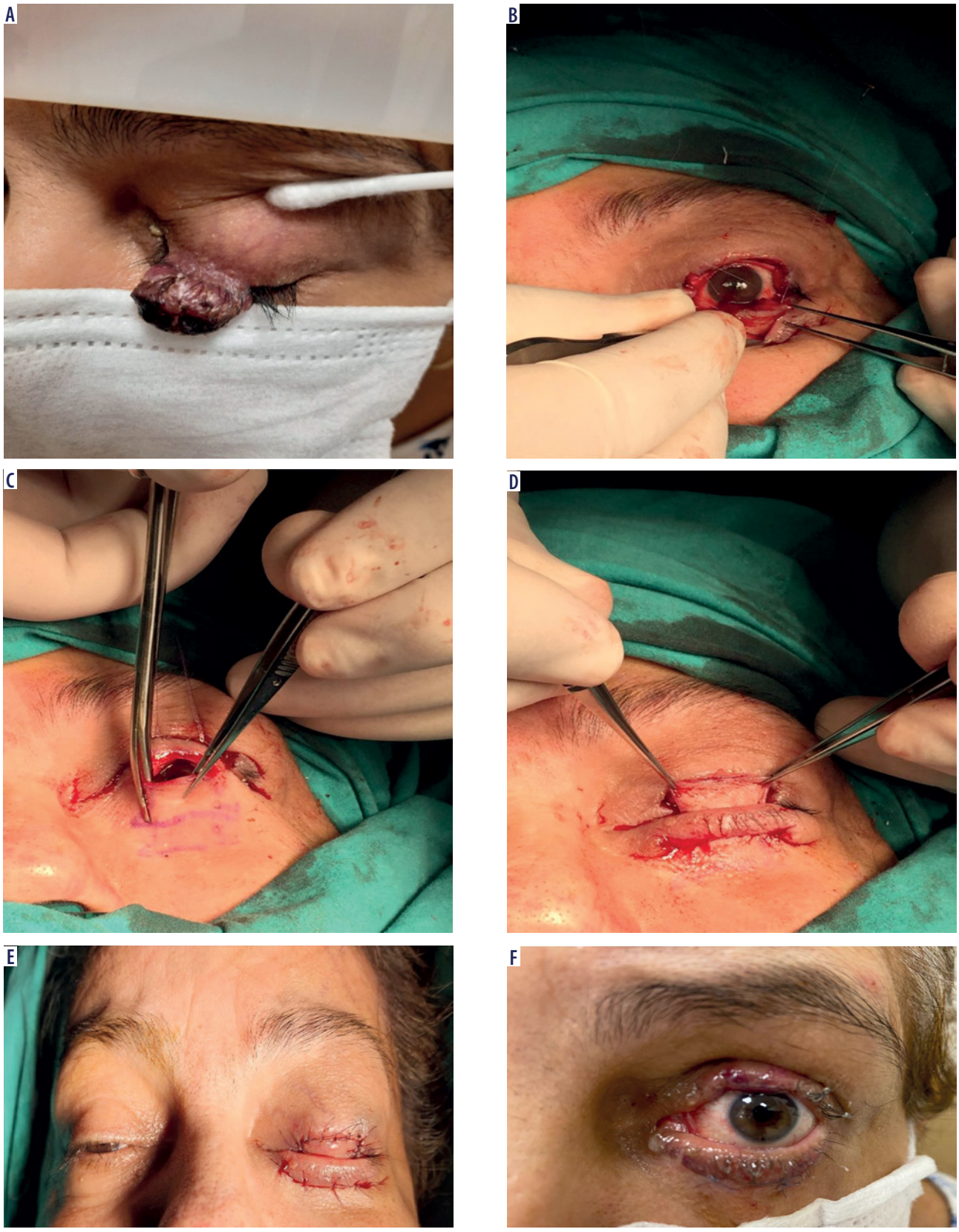

Figure 1. A) Firm and shiny mass with swelling and telangiectasias along the eyelash side on the upper eyelid. B) Large full-thickness upper eyelid defect after tumor excision. C) Making Cutler-Beard flaps. D) Advancing the Cutler-Beard bridge flap under the lower eyelid. E) Suturing the Cutler-Beard flap to cover the defect area. F) Image on the first postoperative week after separating the upper and lower eyelids

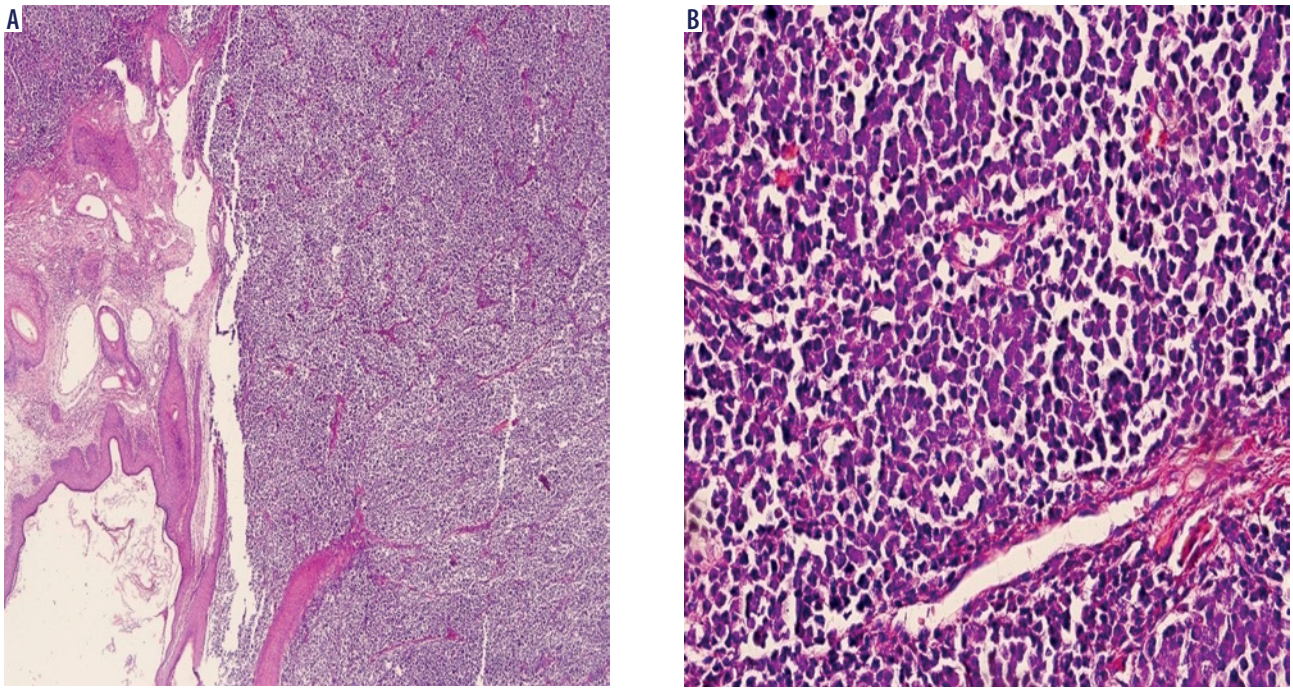

Figure 2. A) Layers filling the dermis under the epidermis, tumor cells with a trabecular or sometimes insular patterns forming nests (H&E, ×40). B) Tumor cells with a neuroendocrine character and a salt-pepper chromatin structure (H&E, ×100)

The patient was diagnosed with MCC and was immediately referred to medical oncology. Systemic imaging and sentinel lymph node (SLN) (parotid, supraclavicular, and jugulodigastric) biopsy revealed no evidence of metastasis. According to the staging system defined by the American Joint Committee on Cancer (AJCC), the tumor was classified as T1N0M0 (< 2 cm, no sentinel lymph node or distant metastasis). The patient was referred to radiation oncology for adjuvant radiotherapy. Because of the location of the tumor, treatment continued in an external center to apply globe-preserving radiotherapy.

Eyelid sutures were removed and lid reconstruction was completed in the 6th postoperative week. The patient was called for a follow-up visit in the first week after the reconstruction. We encountered no early surgical complication (Figure 1). The patient was also planned to attend routine follow-ups for ocular complications due to radiotherapy.

DISCUSSION

Merkel cell carcinoma is a primary cutaneous neuroendocrine carcinoma of the skin [7, 8]. Although rare, it has high risk of local aggressive spread, lymph node involvement, and metastasis and can turn into a life-threatening entity [7, 9].

The most important risk factors for MCC are Merkel cell polyomavirus (MCPyV), UV exposure, immunosuppression and immune escape. MCPyV plays a major role and it has been isolated in 80% of MCC cases [1]. The role of immunosuppression in non-melanoma skin cancers is well known. The risk of developing MCC increases 23-fold compared to baseline after solid organ transplantation [10, 11].

Merkel cell carcinoma can often be mistaken for cysts, chalazion, or basal cell carcinomas and therefore is often misdiagnosed because eye involvement is rare. Although often reported as eyelid involvement, it can also present as conjunctival, lacrimal gland, iris, or orbital metastasis. Merkel cell carcinoma cases have been reported to be nearly twice as high among women compared to men. Compared to other malignant tumors with eyelid involvement, MCC has been reported to be more frequently seen in the upper eyelid. Our case was female and had upper eyelid involvement, supporting the literature.

Our patient had complaints for about 3 months. She had previously been evaluated as having a chalazion in two different centers and had received local antibiotherapy and steroid therapy. The differential diagnosis of eyelid lesions can sometimes prove challenging. Risk factors should be questioned in anamnesis and the definition and invasion status of the mass should be evaluated through careful examination. In particular, lesions with aggressive growth, irregular surface, vascularization, ulceration, and non-healing should be evaluated in favor of malignancy and incisional or excisional biopsy should be performed without delay or hesitation. For malignancies with aggressive growth, as in our case, early diagnosis and treatment significantly increase life span and quality of life.

In cases with suspicion of malignancy, surgical technique is important for the success of functional therapy, aesthetic treatments, and other subsequent treatments. The primary goal should be the complete elimination of the cancerous tissue. The aggressive growth and macroscopic findings of our case suggested that it was primarily malignant. Considering the local aggressive spread and the risk of early metastasis, the Cutler-Beard technique was preferred and wide excision of the tumor was planned. The key advantage of this tech-

nique is that it can be used in most tumor surgical procedures. Particularly for eyelid tumors with planned wide excision, the main goal is to have an accurate and effective surgical technique. Otherwise, complications can occur, including anterior segment problems, upper eyelid entropion, irregularity in the eyelid contour, retraction due to cicatrisation in the lower eyelid or flap necrosis. Also, an incorrect decision for the technique might increase the destructive risks of radiotherapy.

Some studies recommend radiotherapy for MCC cases for the local control of the disease, even when the surgical margin is clean. Adjuvant radiotherapy has been reported to have a positive effect on survival, particularly in primary large tumors [12]. The frequency of use of radiotherapy in MCC cases located in the eyelid varies among centers, although the general belief holds that local radiotherapy should be applied in all MCC patients. For our case, adjuvant radiotherapy was performed to reduce the risk of recurrence and metastasis after surgery [5, 6].

Malignancy should be considered during differential diagnosis for eyelid lesions, particularly in cases with aggressive growth or non-healing. Complete excision of the mass and reconstruction should remain the main goal. For tumors with high risk of distant metastasis such as MCC, early diagnosis, choosing the appropriate surgical technique, and adjuvant treatments have a positive effect on quality of life and survival.

DISCLOSURE

The authors declare no conflict of interest.

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