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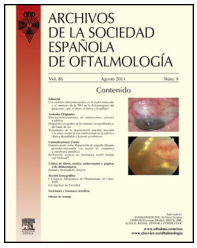

Review

# Merkel cell carcinoma of the eyelid. A series of 5 cases and review of the literature☆

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ABSTRACT

*Objective:* Presentation of 5 clinical cases of Merkel cell carcinoma of the eyelid, highlighting the clinical, histopathological, management, and monitoring features. Review of related literature.

*Methods:* Retrospective review of clinical records and telephone interview of the 5 patients treated for Merkel cell carcinoma between 2006 and 2013, in the Orbit and Oculoplastic Department, Clinical Hospital of the University of Chile.

*Results:* Five patients (2 men, 3 women); 79.2 years (range 64–94 years), with a mean onset of 10 weeks (range 5–16 weeks), tumor size reaching a mean of 2.5 × 2 × 2.1 cm at the time of surgery. Described as a nodular mass, exophytic, solid, reddish colored. With no infiltrated lymph nodes or metastases in the first match. Staging as T2N0M0, after the first surgery with oncological criteria. Two lymph node recurrences detected during monitoring. Distant metastasis was found in one patient. The histopathological diagnosis was confirmed by immunohistochemical study of the biopsy. The surgical strategy was full excision, control of surgical margins in the intraoperative period, and eyelid reconstruction with Hughes flap, Cutler–Beard flap or primary closure, depending on the case.

*Conclusions:* Early diagnosis, wide excision of the tumor with intraoperative control of clear margins with conventional or Mohs surgery, and proper eyelid reconstruction are adequate for a good survival in elderly patients with this eyelid tumor. The study of sentinel lymph node biopsy in primary intervention is recommended; with subsequent radiotherapy to decrease the recurrence and increase survival.



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## Carcinoma palpebral de células de Merkel. Serie de 5 casos y revisión de la literatura

*Palabras clave:*
Células de Merkel
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RESUMEN

*Objetivo:* Estudio descriptivo de 5 casos clínicos de carcinoma de Merkel a nivel palpebral, destacando las características clínicas, histopatológicas, el manejo y seguimiento. Revisión de literatura asociada.

*Métodos:* Revisión retrospectiva de fichas clínicas y entrevista telefónica a 5 pacientes tratados por carcinoma de células de Merkel entre los años 2006–2013, en el Departamento de Órbita y Oculoplástica del Hospital Clínico de la Universidad de Chile.

*Resultados:* Cinco pacientes (2 hombres, 3 mujeres), de 79,2 años (rango 64–94 años), con una evolución de 10 semanas (rango 5–16 semanas), con un tamaño tumoral de 2,5 × 2 × 2,1 cm en promedio al momento de la cirugía. Descritos como una masa nodular, exofítica, sólida, de color rojizo. Sin adenopatías sospechosas ni metástasis en el primer enfrentamiento. Tipificados como T2N0M0, tras la primera cirugía con criterios oncológicos. Se observó, durante el seguimiento, 2 recurrencias en nódulo linfoide. Encontramos metástasis a distancia en un paciente. El diagnóstico histopatológico se confirmó mediante biopsia corriente más inmunohistoquímica. La estrategia quirúrgica fue con extirpación completa, control de márgenes libres y reconstrucción palpebral con colgajo de Hughes, Cutler–Beard o cierre primario.

*Conclusiones:* El diagnóstico precoz, la extirpación amplia del tumor con control de márgenes libres intraoperatorio con cirugía convencional o técnica de Mohs y una adecuada reconstrucción palpebral permiten una supervivencia en pacientes añosos portadores de este tumor a nivel palpebral. Se recomienda el estudio de ganglio centinela y biopsia en la intervención primaria, junto a una radioterapia posterior, que disminuye la recurrencia y aumentaría la supervivencia.



## Introduction

The Merkel cells were described in 1875 by Friedrich Merkel. These cells are found in the skin, specifically in the basal layer of the epidermis, at the dermo-epidermic union. They are also found in the mucosa, except in the conjunctiva.[1] The Merkel cells are neuroendocrine cells and were initially described as tactile sensory cells (mechanoreceptors).[2] At present, these cells are believed to be derived from the neuroectoderm and subsequently migrate toward the skin with the growth of the peripheral nerves.[2,3] In 1972, Tang and Toker described the malignization of these cells, defined at that time as a trabecular carcinoma and at present known as Merkel cell carcinoma or neuroendocrine skin carcinoma.[4] It has been seen that, even though the Merkel cells are in the epidermis, the Merkel cell carcinoma arises in the dermis and extends toward the subcutaneous cell tissue, respecting the overlying epidermis layer.[5] Despite its neuroendocrine nature, no hormonal syndromes associated to the tumor have been reported.[6]

The present descriptive study analyzes 5 clinical Merkel cell carcinoma cases at the palpebral level, in which the clinical, histopathological management and follow-up characteristics are emphasized, in the context of the current findings on the subject.

## Materials and methods

A retrospective review of clinical records and telephone interviews of 5 patients treated for Merkel cell carcinoma between 2006 and 2013 in the Orbit and Oculoplastic Department of the Clinic Hospital of the University of Chile. Information was obtained on the clinic, treatment and subsequent follow-up by the treating team. Informed consents were requested from the 5 patients before publishing the obtained information. Subsequently, the data obtained in monitoring and follow-up of patients were compared with the information drawn from current literature on the subject, obtained from the *PubMed* database.

## Results

Two male and 3 female patients were evaluated, with a mean age of 79.2 years (range 64–94 years). The evolution time since the patients perceived the tumor until their first visit to the ophthalmologist averaged 10 weeks (range 5–16 weeks), with an average tumor size of 2.5 × 2 × 2.1 cm at surgery time. At the macroscopic level, the tumors were described as a nodular, exophytic, solid and reddish mass. One was detailed as an ulcerated lesion on the surface (Fig. 1). One patient had undergone surgery on 2 occasions as chalazia, in a period of