ABSTRACT

Case report: We present the case of a 47-year-old female suffering from progeria who developed an eyelid Merkel cell carcinoma (MCC) following cyclosporine treatment for a corneal transplant. She underwent excision of the lesion by Mohs micrographic surgery (MMS) and received adjuvant treatment with radiotherapy. Later she had a recurrent tumour for which a wide excision with orbital exenteration was performed. The patient was fit and well at the 24-month follow-up.

Discussion: MCC is an aggressive tumour that has a larger incidence in elderly people, women and immunosuppressed patients. Our approach with the patient must be global, avoiding risk factors in predisposed subjects. MMS seems unsuitable for the treatment of MCC; if tumour recurrence occurs a wide resection should be performed (Arch Soc Esp Oftalmol 2009; 84: 581-584).

Key words: Merkel cell carcinoma, Mohs microsurgery, eyelid tumour, orbital exenteration, cyclosporine, young patient.

RESUMEN

Caso clínico: Se presenta el caso de una paciente de 47 años afecta de progeria que después de tratamiento con ciclosporina por trasplante corneal presentó carcinoma de Merkel (MCC) palpebral. Tras exéresis completa mediante cirugía de Mohs (MMS) y tratamiento adyuvante con radioterapia, recidió y fue necesario realizar exenteración orbitaria. La paciente se encuentra asintomática tras 24 meses de seguimiento.

Discusión: El MCC es un tumor agresivo con mayor incidencia en ancianos, mujeres e inmunodeprimidos. Nuestra visión del enfermo debe ser global evitando añadir factores de riesgo a pacientes ya predisuestos. La MMS parece desaconsejada en el tratamiento del MCC, debiendo realizar una exéresis radical si se produce recidiva.

Palabras clave: Carcinoma de Merkel, microcirugía de Mohs, tumor palpebral, exenteración orbitaria, ciclosporina, paciente joven.
INTRODUCTION

The Merkel cell carcinoma (MCC) is a highly invasive cutaneous neoplasia. Several treatment methods have been postulated, including local excision with broad margins, Mohs’ micrographic surgery (MMS) with or without radiotherapy, and radical excision with orbitary exenteration (1).

CLINICAL CASE

A 47 year-old Caucasian female affected with progeria with bone malformations (pectus excavatum, malar bones hypoplasia and zigomatic arches), visceral malformations (hepatomegalia, dilatation of the left kidney pelvis), ferropenic anemia, venous insufficiency, antecedents of erysipelas and classic Kaposi’s sarcoma in the distal third of both lower limbs, treated with interferon ten years back. She have been intervened for implanting a prosthesis for malar bones, mammoplasty, several endometrial curettages and appendectomy. Her parents were cousins. At the ocular level she exhibited myopia magnus, cataracts intervention in both eyes, two pterigium interventions in right eye with residual corneal leukoma, which led to a penetrating keratoplasty in 2002. The tissue was rejected and it was decided to initiate immunosuppression with cyclosporine, receiving a new corneal transplant. In the hospital where the transplants were made a nodular purplish lesion in the upper right eyelid was biopsied in late 2005. The lesion had appeared three months earlier and in the last two had increased its growth rate. It was reported as MCC (fig. 1), at which point the patient was referred to our hospital.

The exploration of the entire body skin, palpation of ganglionary chains and computerized axial tomography (CAT) of cranium-chest-abdomen were normal, which led to classifying the tumor as a localized disease.
In December 2005 the tumor was removed by means of two-stage MMS (fig. 2). The lymphography produced a diffuse capture of the latero-cervical ganglionary chain which was a contraindication for the sentinel ganglion. The patient was treated with radiotherapy on the primary tumor area and right latero-cervical ganglionary chains with 50 Gy (total dose). Four months later three hard nodules of 2 cm each appeared (fig. 3A) in the right upper and lower eyelids and temporal region of the same side which, once biopsied, were reported as MCC.

It was decided to intervene with a right side orbitary exenteration enlarged to the homolateral temporal region, leaving margins of 2.5 cm from the lesions (fig. 3B). The orbit was covered with a fronto-temporal rotated miocutaneous flap (figs. 3C and 3D) and the remaining defect was covered with a total skin graft from the left supra-clavicular region (figs. 3E and 3F). The immediate post-op did not exhibit complications (fig. 4).

Subsequent controls were negative and after 24 months of follow-up, no relapses have emerged (fig. 5).

**DISCUSSION**

MCC is a rare neuroendocrine skin neoplasia first described by Toker in 1972 (1,2). Typically it appears in elderly patients [75% are 65 or more (2)],...
although it has also been described in young adults with congenital ectodermic dysplasia syndromes.

Its prevalence is increased in areas exposed to the sun and after immunosuppression and organ transplants. It is more frequent in white-skinned individuals and twice more frequent in women. Half of MCC’s appear in the head and neck region and one tenth in eyelids or periocular region with a preference for the upper eyelid.

The instant case is a patient with a poly-malformation syndrome and early aging as demonstrated by the presence of cataracts, pterygium and Kaposi’s sarcoma. In this context, cyclosporine therapy should have been avoided for an immunodepressed patient. We cannot state that the MCC appeared as a consequence of said treatment but the Kaposi sarcoma is a clear contraindication for this medication. This reminds us that we must study the patient globally and not only from the viewpoint of our specialty.

MCC is aggressive and has a high local recurrence rate (30% at one year), lymphatic invasion (50-60% of cases) and metastasis (over 30% of cases) (1,2). The 5-year survival rate of cutaneous MCC is 38% (1,2), although some spontaneous regression cases have been described (2).

The treatment for localized MCC is surgical, with 2.5 cm margin surgery proposed (3). MMS respects the largest possible amount of tissue because, during the operation, it studies all the tumor edges (4). However, this technique does not seem adequate for treating Merkel carcinoma, although some cases of healing have been published (5). In other studies it is stated that the cold cuts do not allow an adequate study of these tumoral cells and even though Mohs surgery associated with radiotherapy seems to produce better results (5) we do not consider it to be an adequate technique for this tumor. For local MCC relapses we must perform radical surgery because healing is possible (2).

Orbital exenteration and reconstruction with frontotemporal miocutaneous graft is a simple technique which allows vascularization of the anophthalmic cavity with a fine thickness which facilitates the detection of relapses and achieves an acceptable esthetic result.

REFERENCES