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SHORT COMMUNICATION

Case report

Merkel cell carcinoma of the eyebrow extending into the orbit

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BACKGROUND. *Merkel cell carcinoma is a rare and aggressive cutaneous neoplasm with high local recurrence and metastatic rates.*
PURPOSE. *To highlight an unusual presentation of a Merkel cell carcinoma.*
CASE REPORT. *We report a large Merkel cell tumor in an 85-year-old woman. It extended from the eyebrow into the upper nasal orbit posterior to the equator of the eye globe. Local resection resulted in microscopically complete removal of the tumor. During follow-up (20 months) no local recurrence was noticed but there was a metastasis in a lymphatic nodule.*
CONCLUSIONS. *Merkel cell carcinoma often presents in the eyelids and periorbital region. However, extension from the eyebrow deep into the orbit is unusual. (Eur J Ophthalmol 2000; 10: 330-1)*

KEY WORDS. *Merkel cell tumor, Eyelid, Orbit, Carcinoma*

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INTRODUCTION

Merkel cell carcinoma is a primary cutaneous tumor which originates from precursor cells of Merkel cells. These tumors are often found in the eyelids and periocular region (1-3) but extension into the orbit is rare (1). We describe a case of Merkel cell carcinoma of the eyebrow extending deep into the orbit.

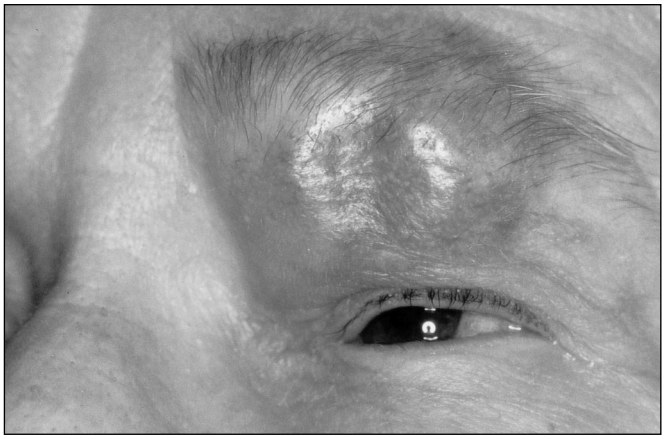

Fig. 1 - *Merkel cell carcinoma beneath the left eyebrow in an 85-year-old woman. Firm protuberant lesion with livid color and typical teleangiectatic blood vessels at the surface.*

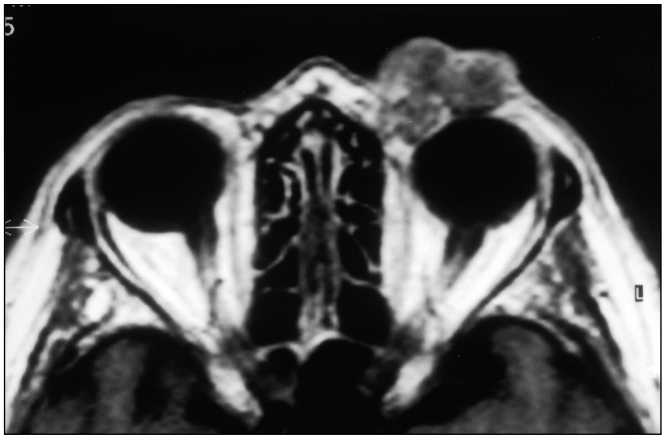

Fig. 2 - *MRI scan showing extension of the tumor from the eyebrow into the upper nasal orbit to the equator of the eye globe.*

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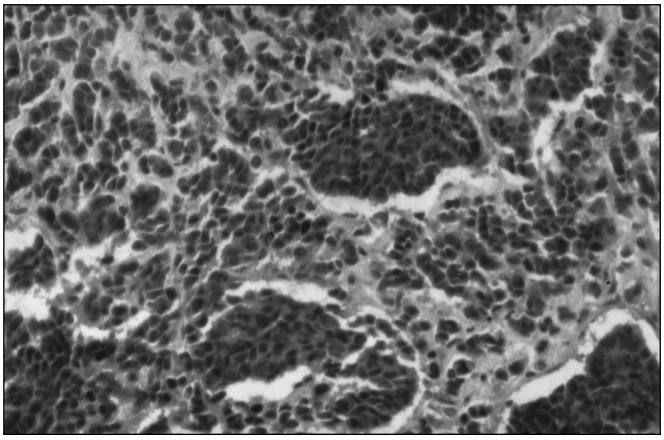

Fig. 3 - *Merkel cell carcinoma with medium-sized cells, scanty cytoplasm and isomorphous nuclei (neuron-specific enolase staining, 250x)*

Case report

An 85-year-old woman had an approximately six-month history of an expanding nodule beneath the left eyebrow. She was otherwise in good health. On examination, there was a firm dermal tumor (3.1 x 2.2 cm) of livid color beneath the left eyebrow, tightly adhering to the overlying epidermis (Fig. 1). The tumor extended into the upper nasal orbit to the equator of the eye globe (Fig. 2). There was no lymphadenopathy of the head or cervical region. Eyelid opening was incomplete but the motility of the globe was not disturbed. Local resection resulted in microscopically complete removal of the tumor. Histopathologic findings were consistent with Merkel cell carcinoma (Fig. 3). During follow-up (20 months) no local recurrence was noticed. A metastasis occurred in a parotid lymph node. Extirpation of the metastasis was followed by irradiation therapy (60 Gy).

DISCUSSION

Merkel cell carcinoma probably develops from the precursor cells of Merkel cells that are found in the epidermis and outer root sheaths of hair follicles. They share ultrastructural features with neuroendocrine cells (1). The diagnosis of Merkel cell carcinoma is based on features typical of normal Merkel cells. Typical electron microscopic findings are dense core granules, intranuclear rodlets and spinous processes. Antigenic features include a positive reaction for specific cytokeratins, epithelial membrane antigen, neuron-specific enolase (Fig. 3), chromogranin and synaptophysin (1). The tumor often mimics lymphoma or undifferentiated carcinoma (4).

Merkel cell tumors tend to be bulging lesions near the lid margin of elderly patients. They are reddish and erythematous with teleangiectatic vessels (2, 3). The clinical diagnosis is often difficult and misleading (2). Almost 1 out of 10 Merkel cell tumors affects either the eyelid or the periocular region (1). In our patient a large Merkel cell tumor extended from the eyebrow deeply into the orbit; this presentation is very unusual (1).

Merkel cell carcinoma is a rare and aggressive tumor. One third of Merkel cell carcinomas recur, and almost two thirds give rise to regional node metastases, as in our patient. Up to one half of Merkel tumors metastasize widely and result in death. Initial treatment should be prompt and aggressive, with wide resection and routine postoperative irradiation. Although metastatic lesions often respond to radiation therapy and cytostatic drugs, these treatments are mainly of palliative value.

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