

SHORT COMMUNICATION**Case report****Merkel cell carcinoma of the eyelid: a case report**L. COLLAÇO¹, J.P. SILVA^{1,2}, M. GONÇALVES³, P. ABRANTES¹¹ Department of Ophthalmology, Hospital de S. José, Lisboa² Department of Cellular Biology, Faculdade de Ciências Médicas de Lisboa, Lisboa³ Department of Pathology, Hospital de S. José, Lisboa - Portugal

A 22-year-old woman who presented with a tumour of the upper eyelid with clinical, histological, immunohistochemical and ultrastructural features of a Merkel cell carcinoma is reported. This is a unique case of Merkel cell carcinoma in such a young patient. This neuroendocrine tumour is typically found in the elderly. It can grow rapidly, has the potential for local recurrence and early metastatic spread. The authors recommend wide resection of the primary site, which proved effective in this case. (Eur J Ophthalmol 2000; 10: 173-6)

KEY WORDS. *Eyelid, Merkel cell, Carcinoma*

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INTRODUCTION

The Merkel cell is an epidermal cell with distinct ultrastructural characteristics. At the periphery it contains dense-core cytoplasm granules and filaments, which are features of cells in the diffuse neuroendocrine system (1). A new, distinct entity was first described in 1972 by Toker that he called trabecular carcinoma of the skin (2). Speculation that these tumours arose from Merkel's cells in the skin began in 1978, when an ultrastructural study by Tang and Toker demonstrated the presence of neurosecretory granules in these trabecular carcinomas (3). Further studies supported this concept and the name "Merkel cell carcinoma" became commonly used (4-6).

Head and neck are the typical sites of Merkel cell neoplasms but these lesions have seldom been described on the eyelid (7-9). This report describes a Merkel cell carcinoma of the eyelid in a young patient, although the tumour is usually found in elderly persons.

Case Report

A 22-year-old woman presented with a five-week

history of a progressively enlarging, nontender nodule on the right upper eyelid (Fig. 1). It was well-circumscribed, reddish-purple, 15 mm in diameter, and protuberant, with visible superficial vessels. Blood chemistry studies showed no abnormalities. Wide local excision was performed and pathological examination was done. We dissected two segments. One was immediately placed in 10% neutral buffered formaldehyde, and prepared for paraffin histology using standard techniques. Sections of 3 µm were cut by a Reichert microtome for histology (hematoxylin-eosin and Gomori) and immunohistochemistry (neuron-specific enolase of the cytoplasm granules – NSE, and low-molecular-weight-cytokeratin-CK).

Histological examination showed anastomosing cords and nests of cells of uniform size and shape. They had round nuclei and scanty, ill-defined cytoplasm (Fig. 2). Immunohistochemistry (Figs. 3, 4) showed positive staining for NSE of the cytoplasm granules and low-molecular-weight CK.

The second segment was placed in 2.5% glutaraldehyde, post-fixed in 1% osmium tetroxide, rinsed in cacodylate buffer, and dehydrated in a graded series of alcohols. The specimens destined for transmission microscopy were embedded in epoxy resin; 1 µm-thick sections were made with toluidine blue, and 60 nm thick sections were stained with aqueous uranyl acetate and

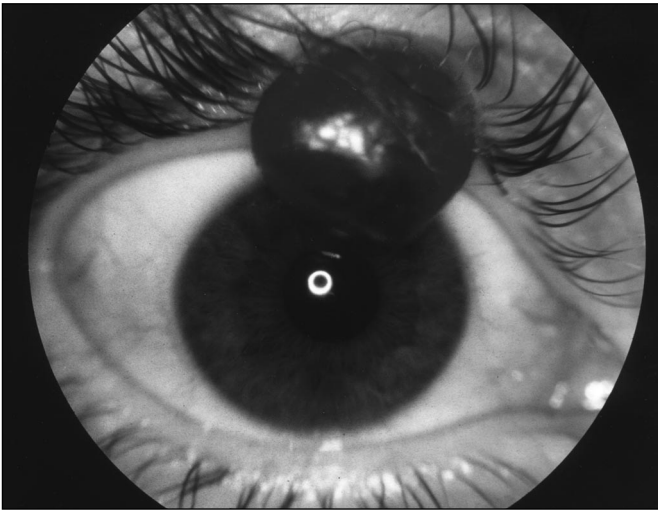


Fig. 1 - *Violaceous nodule on the upper eyelid.*

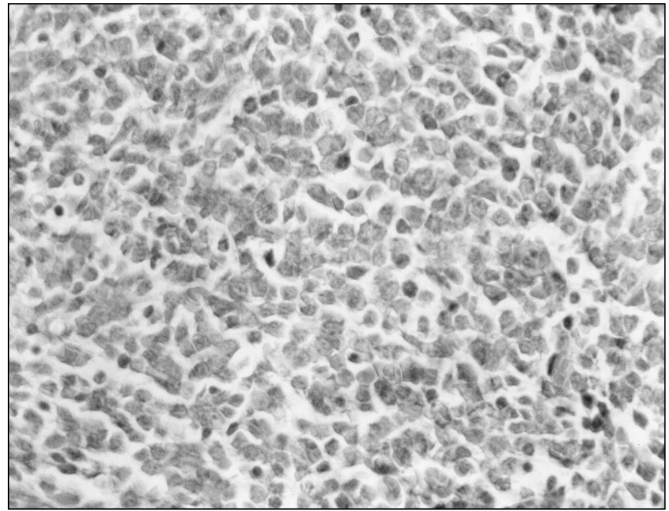


Fig. 2 - *Light microscopy: cytologically uniform cells have round nuclei and scanty ill-defined cytoplasm (HE 100x).*

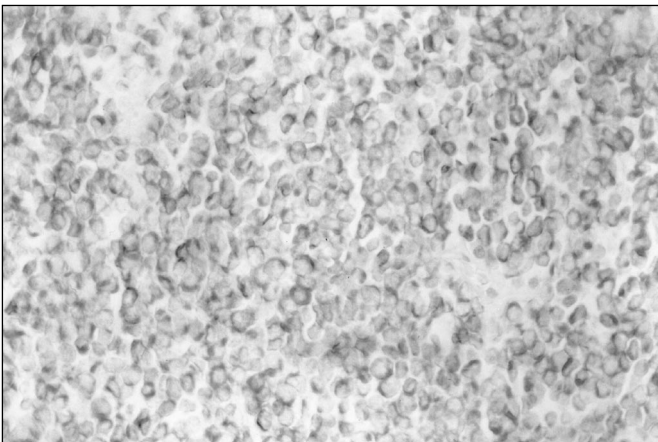


Fig. 3 - *Immunohistochemistry: positive staining for low-molecular-weight cytokeratin (CK 400x).*

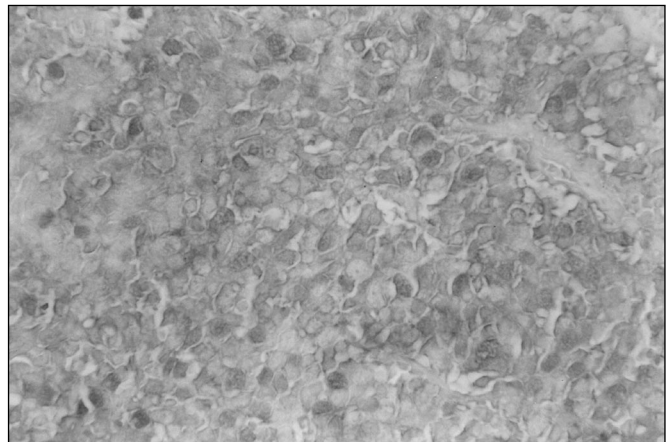


Fig. 4 - *Immunohistochemistry: positive staining for neuron-specific enolase of the cytoplasmic granules (NSE 400x).*

lead citrate. The sections were examined with a JEOL JEM-100CXII transmission electron microscope.

Electron microscopy showed polygonal tumour cells with central nucleus. There were no desmosomes between tumour cells. The cytoplasm was ill-defined and contained dense-core neuro-secretory granules. These were close to the nucleus and aligned along the cell membrane (Fig. 5). The histological and ultrastructural characteristics of this eyelid neoplasm were typical of Merkel cell carcinoma. No evidence of local recurrence or metastatic spread has been detected three years after excision.

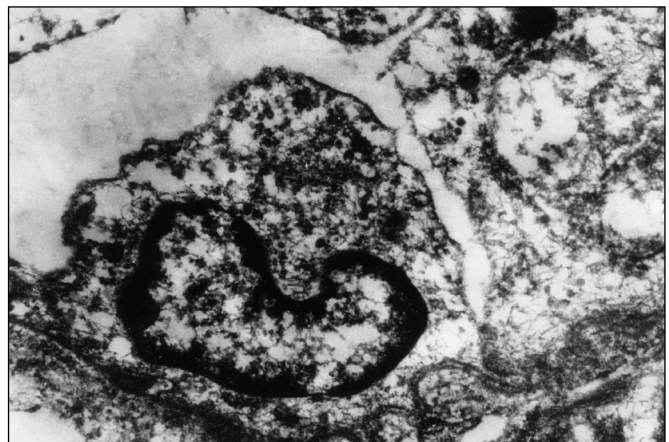


Fig. 5 - *Electron microscopy: intra-cytoplasmic neuro-secretory granules (14000x).*

DISCUSSION

Merkel cell carcinoma was first described by Toker in 1972 but its precise origin is still uncertain (2). It is a neuro-endocrine tumour that most often appears on sun-exposed skin, typically in people aged 60 or more (mean age 68.8 years), both sexes being equally affected (8). Unusually, our patient was only 22 years-old.

Clinically, the tumour has been described as a single firm nodule with blue, red or violaceous overlying skin, sometimes with very fast growth (5, 8, 10-12). The differential diagnosis includes other primary cutaneous tumours, metastatic deposits of oat cell carcinoma and lymphoblastic lymphoma (6, 8, 11, 13, 14).

Histologically, the tumour shows a trabecular and nest-like pattern. The cells have a uniform appearance with pale empty nuclei and scanty pale cytoplasm. Immunohistochemistry has proved helpful in diagnosis, positive staining for NSE and for CK being strongly suggestive of Merkel cell carcinoma. Ultrastructurally, the cytoplasm contains dense-core neuro-secretory granules and perinuclear whorls of intermediate filaments (5, 8, 10).

Merkel cell carcinoma is an aggressive tumour with a recurrence rate of approximately 40% and

an incidence of distant metastases of about 30% (14). The estimated five-year survival rate is 38% (5, 10, 15). That is why it is important to make a prompt diagnosis. This can be done confidently with the aid of immunohistochemistry and electron microscopy.

The best mode of therapy is still unclear but, like other authors (10, 14, 16, 17), we treated the tumour with wide surgical resection alone. We would reserve the use of radiotherapy for tumour recurrences or regional lymph node spread, and chemotherapeutic regimens for systemic metastases.

We emphasise the need for histological diagnosis of tumour-like lesions of the eyelid and recommend wide surgical resection for patients with this tumour.

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