CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 7 (2015) 104–108

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Spontaneous regression of Merkel cell carcinoma: A case report and review of the literature

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ARTICLE INFO

Article history:
Received 29 July 2014
Received in revised form 7 November 2014
Accepted 8 November 2014
Available online 13 November 2014

Keywords:
Merkel cell carcinoma
Spontaneous regression
Nose
Head and Neck
Skin
Cancer

ABSTRACT

INTRODUCTION: Merkel cell carcinoma (MCC) is a rare and highly aggressive primary cutaneous neuroendocrine carcinoma, most often occurring in the elderly. Recurrence is frequent and in 40% of cases regional and distant metastases develop. Despite this, there have been reports of spontaneous regression. We report the first case of MCC with primary complete spontaneous regression of the nose in an 86-year-old woman following an incisional biopsy.

PRESENTATION OF CASE: An 86-year-old woman presented with a violaceous lump on the left side of the nose measuring 25 × 25 mm. Incisional biopsy of the lesion showed MCC and immunohistochemistry confirmed diagnosis. Following an 8-week period the lesion completely disappeared and histology did not show any residual MCC but immunohistochemistry demonstrated a mixture of T and B cells.

DISCUSSION: Complete spontaneous regression (CSR) is rare. The literature documents 22 similar cases of CSR of MCC. From this case report and previous literature the most likely reason for regression is a T-cell mediated immune response.

CONCLUSION: To the best of our knowledge, this is the first described case of MCC with primary CSR of the nose. Exact mechanism of regression remains unclear. Further research is needed in identifying pathway of immune response and possible immunotherapy as a cure.

1. Introduction

Merkel cells are primarily located in the basal layer of the epidermis and concentrated in touch-sensitive areas of the skin.[1] Their most noticeable ultrastructural characteristics are the dense-core secretory granules accumulated near the nerve fibre junction, which may contribute to its indefinite neuroendocrine function.[2]

Merkel cell carcinoma (MCC) was first described by Toker[3] in 1972 as trabecular carcinoma of the skin. 85% of all MCCs appear on sun-exposed areas[4] with the head and neck region most frequently affected, accounting for 35–47% of these cases.[5,6] The prognosis is poor, with a 5-year survival rate of around 60%[7], owing to the common involvement of regional lymph nodes (10–45%) at initial presentation, of which 50–75% of patients develop regional lymph node metastases at some time.[8–12] Distant metastases commonly affects 50% of patients with common sites being the lymph nodes, liver, bone, brain, lung and skin.[8–10,13,14]

O'Rouke and Bell[15] first described complete spontaneous regression (CSR) of MCC in 1986. Thereafter, 21 additional cases have been reported. This present study presents a case of complete spontaneous regression of MCC with an immunohistochemistry study of the region in which the tumour was located.

2. Presentation of case

An 86-year-old female patient presented with a violaceous lump on the left side of the nose measuring 25 × 25 mm. An incisional biopsy was performed and histology showed a dense infiltrate of small tumour cells with hyperchromatic nuclei and little cytoplasm (Fig. 1). Immunohistochemistry confirmed the diagnosis of MCC, with positive staining for cytokeratin 20 (CK20) (Fig. 2), neuron-specific enolase (Fig. 3), synaptophysin and negative staining for cytokeratin 5/6T, TTF1 and MelanA. The patient attended for an excision 8 weeks after her initial biopsy and showed no presence of lump prior to excision (Fig. 4). Histology of the excised specimen showed severely sun-damaged skin with mild epidermal atrophy. There was a patchy chronic inflammatory cell infiltrate with focal fibrosis and foreign body giant cell reaction with no evidence of any residual MCC. Immunohistochemistry demonstrated a mixture of both T and B cells with positive staining for CD4, CD3, CD5, and

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http://dx.doi.org/10.1016/j.ijscr.2014.11.027

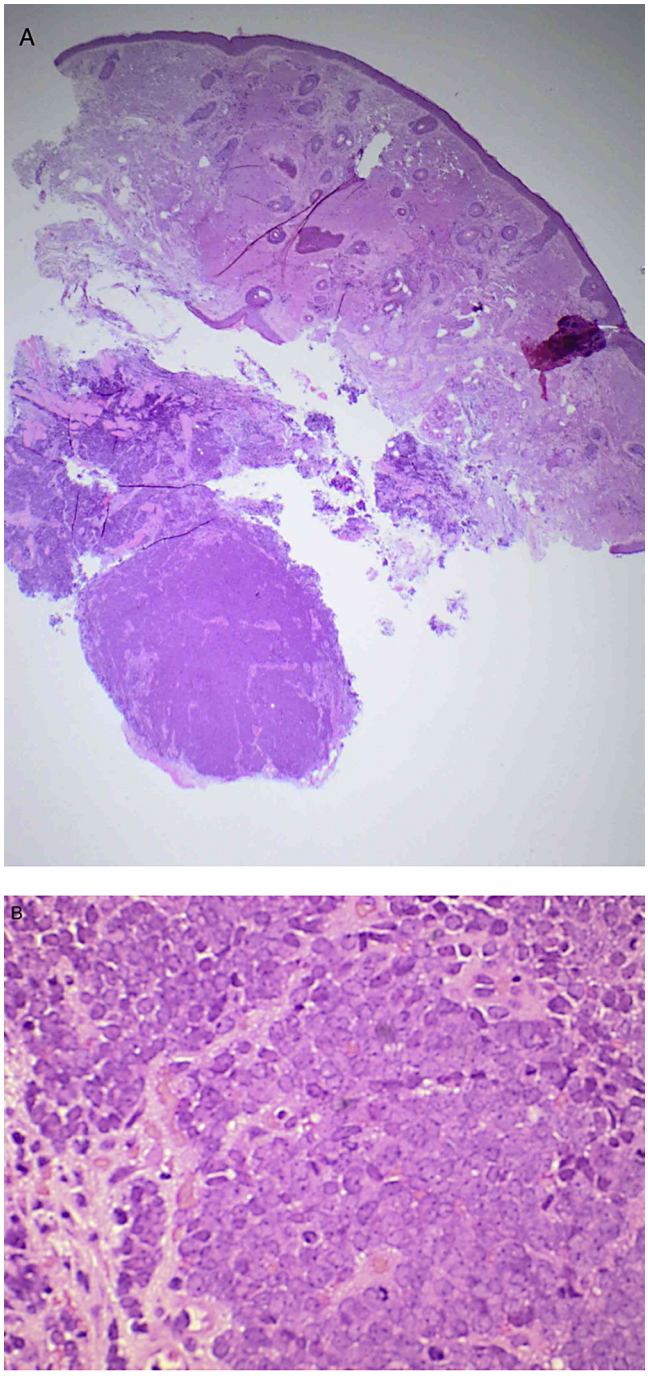

Fig. 1. Histopathological examination showing dense infiltrate of small tumour cells with hyperchromatic nuclei and little cytoplasm. (A) Haematoxylin-eosin, magnification 20×. (B) Haematoxylin-eosin, magnification 400×.

CD8 with CD4 slightly more than CD20 and CD79a. In addition, the specimen revealed admixed macrophages, which stained with CD68.

3. Discussion

Spontaneous regression among all neoplastic diseases has been estimated to be 1 case per 60,000 to 100,000 neoplasms.[16] Complete spontaneous regression (CSR) of MCC is rare and predicted to be 0.0013%.[17] To date, 15 cases of complete MCC regression following incisional biopsy have been reported (Table 1), along with 7 cases of regression occurring after local or regional recurrence of

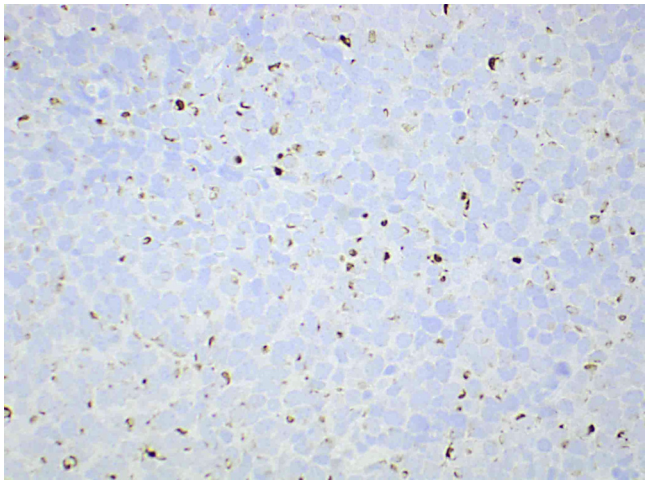

Fig. 2. Positive immunostaining for cytokeratin 20 showing a dot-like pattern (magnification 400×).

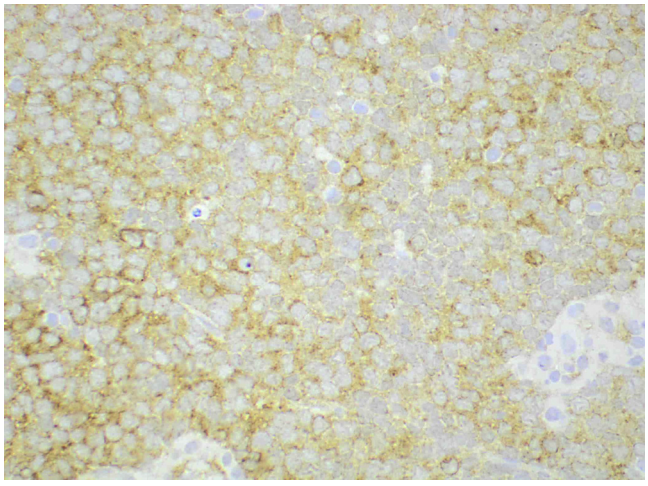

Fig. 3. Positive immunostaining for neurone-specific enloase (magnification 400×).

the carcinoma (Table 2). In the group of MCCs with primary CSR most neoplasms were located on the cheek. In contrast, none of the cases of MCCs with CSR after local recurrence or metastasis were primarily located on the cheek. In both groups of patients the majority were female (15 cases) and the mean age was 79 years old. Our patient presented the typical characteristics of patients with primary CSR of MCC i.e. female sex, elderly and regression after incisional biopsy.

The histopathologic study of the biopsy following CSR in our patient demonstrated results similar to other reports.[18–20] The biopsy showed accumulation of chronic inflammatory cells, mainly T cells. Other studies of both primary and secondary MCC demonstrated infiltration by CD4+, CD8+ and CD3+ T lymphocytes and foamy macrophages.[17–21] The mechanism of CSR remains unclear, however, along with our findings, it suggests that T-cell-mediated immunity plays an important role in tumour regression. This could be attributed to the initial incisional biopsy of MCC (15 cases), which may have triggered tumour regression via stimulation of the immune system. In addition, the majority of patients were elderly with poor health status and various co-morbidities, which may suggest other unknown mechanisms, could be involved.