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Merkel cell carcinoma in East Yorkshire: A case series and literature review of current management

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Summary *Introduction:* Merkel cell carcinoma (MCC) is a rare, aggressive neuroendocrine tumour of the skin. The incidence is rising and it is associated with sun exposure and immunosuppression. Our aim was to perform a 10-year retrospective review of MCC treated in East Yorkshire and to examine disease progression, surgical and adjuvant management, and outcomes.
Methods: A 10-year retrospective review was undertaken of patients identified through the histopathology database. Case notes and digital patient records were examined for patient demographics, disease characteristics, management and outcome. Disease stage was calculated using the 2010 AJCC TNM classification.
Results: Thirty-seven patients with complete records were included. Twenty-one patients were male and 16 female, with mean age 76.7 years at presentation. Pre-malignant or malignant skin changes were documented in 15 patients, and immunosuppression in 15 patients. Mean duration of lesion was 17.5 weeks. Following diagnosis 22/37 patients underwent further surgery with 11 patients undergoing sentinel lymph node (LN) biopsy. LN disease was palpable at presentation in 8 patients. Three year survival is 40%.
Conclusions: There is no standardised management of MCC and randomised trials are challenging due to relatively small numbers. There has been little progress made in terms of improving survival. Development of a national database for patients with this condition would

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allow prospective data collection and more accurate assessment of current treatment protocols and their efficacy.

Level of Evidence: IV

Introduction

Merkel cell carcinoma (MCC) is a rare neuroendocrine tumour of the skin. Originally described by Toker in 1972[1] it was initially thought to be an indolent disease but experience of this malignancy has demonstrated it to be a highly aggressive process. The natural history of the disease is predictable with spread via lymphatics before metastatic disease presents. The reported incidence of MCC is on the rise[2] and this is likely multifactorial. MCC typically affects older generations and as the population ages skin malignancies associated with increasing age, immunosuppression and lifetime sun exposure are increasing. In particular MCC has demonstrated a significant increase in age-adapted incidence.[3] Other contributing factors to a rising incidence include improved pathological examination and diagnosis, and a higher index of suspicion for skin cancer in general. Recently the discovery of Merkel cell polyomavirus infection (MCPyV) has been linked to MCC and the role of this infection and possible influence are under investigation.[4,5] Treatment of MCC is a controversial issue and includes surgery, radiotherapy and chemotherapy.

In East Yorkshire the Plastic Surgery department undertakes the management of patients diagnosed with MCC. There is currently no clear consensus locally, nationally or internationally on the best practice for managing these patients. As a result we decided to analyse our local population who have MCC, examine our treatment and outcomes, and determine whether we could draw any conclusions on best management.

Methods

Patients with a diagnosis of MCC between 2001 and 2011 were identified through a database kept prospectively by the histopathology department at Hull and East Yorkshire (HEY) NHS Trust. This search included those patients whose diagnosis was made in another hospital and their details transferred to HEY NHS Trust for discussion at the multidisciplinary team meeting. A retrospective review of the notes was performed. Data collected included basic patient demographic details, past medical history, tumour characteristics and treatment undertaken including surgical and adjuvant therapies.

The tumours were staged according to the American Joint Committee on Cancer (AJCC) Staging Manual 2010.[2]

Data is presented descriptively: The mean (sd) for continuous data and n (%) for categorical data. Kaplan Meier survival curves were drawn for time from diagnosis to death or last follow-up, with a, log rank test to compare stage at presentation. All analyses were undertaken on SPSS (v19).

Results

Patient demographics and tumour characteristics

Forty-two patients were identified through the database as having a diagnosis of MCC. Five patients were excluded due to unavailability of their notes for review or incomplete documentation. Therefore 37 patients were included of which 21 were male and 16 were female, the mean age at presentation was 76.7 years (median 79 years, range 53—93 years). In 34 patients the median duration of the lesion prior to presentation was 12 weeks (range 4—104 weeks). Location of the primary tumours were as follows — head and neck 17 (46%), upper limb 7 (19%), trunk 3 (8%), lower limb 10 (27%) (Figure 1). Treatment was undertaken by the Plastic Surgery department in 84% of cases (31 patients), with the remainder of cases under the care of Dermatology (3 patients), Otolaryngology (2 patients) and Maxillofacial surgeons (1 patient). Fifteen patients (40.5%) were noted to have or have had a diagnosis of a skin malignancy (BCC or SCC, n = 10) or a predisposing skin condition (Bowen's disease or actinic keratosis n = 5) prior to or concurrently with the diagnosis of MCC. Eleven patients (30%) were identified as being immune-compromised due to haematological malignancy, other primary organ malignancy and immunosuppression following organ transplant. Disease stage at presentation is shown in Figure 2. Stage I disease was diagnosed in 40.5% of patients at presentation, stage II disease in 27%, stage III disease in 21.5%, and stage IV disease in 8% of patients. Staging was not possible in one patient whose tumour size was not commented upon.

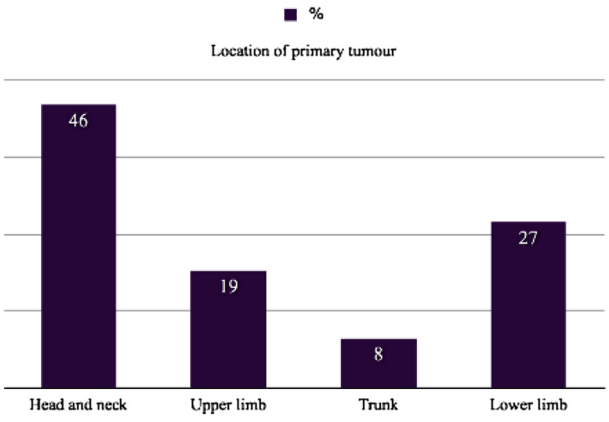

Figure 1 Location of primary tumour.