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Clinical case

Merkel cell carcinoma of the hand: Case report and literature review

Tumeur de Merkel de la main : à propos d'un cas et revue de la littérature

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Abstract

We are reporting on a 72-year-old male who was diagnosed with Merkel cell carcinoma on the dorsal aspect of his left index finger. This rare highly aggressive malignancy of the skin has only exceptionally been described on the finger or hand. This case report helps review important findings associated with this rare malignancy and reviews the pertinent literature.

Keywords: Hand; Merkel cell carcinoma; Ray amputation; Aggressive; Mortality

Résumé

Les tumeurs de Merkel sont des tumeurs rares et agressives dont la localisation aux extrémités est exceptionnelle. Avec seulement deux cas dans la littérature, sa prise en charge thérapeutique n'est pas bien définie. Nous présentons notre prise en charge d'une tumeur de Merkel de la face dorsale de l'interphalangienne distale de l'index d'un l'homme de 72 ans ayant bénéficié d'une amputation du second rayon et faisons un état des lieux du traitement de cette lésion.

Mots clés : Main ; Tumeur de Merkel ; Amputation du rayon ; Agressif ; Mortalité

1. Introduction

Toker reported trabecular carcinoma or Merkel cell carcinoma (MCC) for the first time in 1972. MCC is an uncommon and aggressive malignancy of the skin. Typically patients present with a rapidly growing, firm, non-tender cutaneous nodule, with a red or bluish color. The nodule can measure up to several centimeters in its greatest dimension.

Diagnosis is usually made following biopsy, and even then diagnosis is difficult because of its undifferentiated histological appearance. There is no consensus regarding the optimal therapeutic approach, with relative roles of surgery, radiotherapy, and chemotherapy are still controversial. The rare presence of this rare tumor on the finger or hand adds more controversy on its treatment.

2. Case report

A 72-year-old Caucasian white male was referred for treatment of a mass on the dorsal aspect of the proximal interphalangeal joint of his left index finger. The nodule had been present for 10 years or more with no significant changes in dimensions but occasional episodes of spontaneous bleeding had been reported. Past medical history included diabetes mellitus, hypertension and hypercholesterolemia, gout, and Dupuytren's disease on his left hand. He had also been treated for multiple basal cell carcinomas on the face.

General clinical examination was unremarkable. Local examination showed a 1 × 1 cm nodule on the dorsal aspect of his left index finger (Fig. 1). The mass was firm, slightly bluish, and non-tender.

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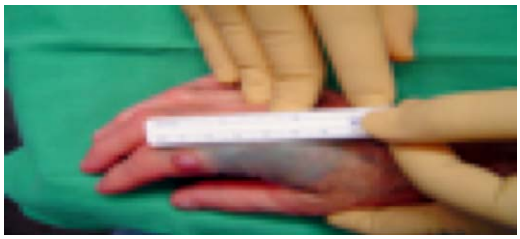

Fig. 1. 1 × 1 cm nodule on the dorsal aspect of left index finger.

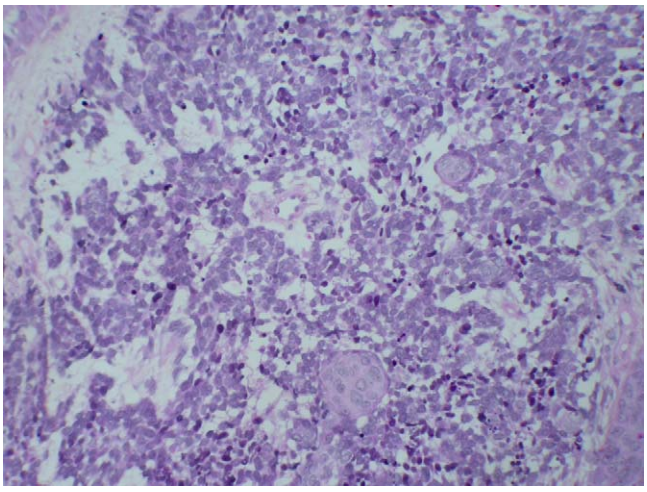

Fig. 2. HPS stain ×20, Anastomosing nets of round cells.

Patient was seen by a surgical oncologist. No regional lymph nodes were palpable. A biopsy was performed and the diagnosis of MCC of the hand was made. A second opinion from a specialized dermatopathologist confirmed this rare diagnosis using immunohistochemistry (Figs. 2–4). Chest X-ray was performed to rule out a small cell carcinoma of the lung. Ultrasound examination of the liver was normal. Surgery was scheduled. Two weeks before surgery, the patient reported that the lesion had progressed in size. Indeed, examination of the lesion the day of the surgery showed increase in size to about 2 cm.

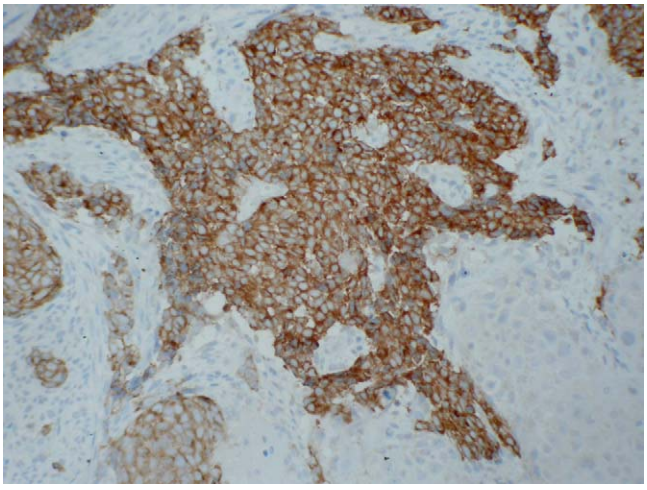

Fig. 3. Cytoplasmic staining synaptophysin – ABC ×100.

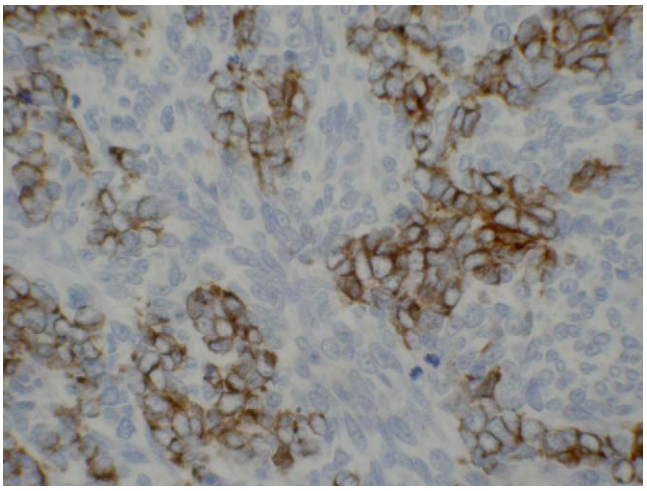

Fig. 4. Keratin 20 (Paranuclear dot) – ABC ×80.

A ray amputation of the left index finger was performed in addition to a sentinel lymph node biopsy. Surgical margins following amputation measured at least 2.5 cm from the tumor margins.

Pathology confirmed the initial diagnosis of MCC. Sentinel node biopsy was performed using intradermal injections of Technetium sulfur colloid and lymphazurin. Five biopsied sentinel nodes were found free of tumor. The wounds healed uneventfully and hand therapy was prescribed. Patient was referred to the radiation oncology department and the decision was taken not to give any adjuvant radiation therapy based on the fact that ray amputation was performed with 2.5 cm margins and no at-risk-of-recurrence wound area existed to irradiate. The patient was seen 24 months postoperatively with no clinical evidence of any recurrence of the tumor.

3. Discussion

Since it was first reported in 1972 by Toker [1], this tumor has been referred to by a variety of names including cutaneous amine precursor uptake and decarboxylation (APUDoma), primary small cell carcinoma of the skin, neuroendocrine carcinoma, primary undifferentiated carcinoma of the skin, malignant Merkel cell tumor, primary small cell carcinoma of the skin with endocrine differentiation, and anaplastic carcinoma of the skin. Merkel cell carcinoma is the most common name for this skin cancer because of the presence of neurosecretory granules within tumor cells.

MCC is an uncommon aggressive malignancy of the skin with only about 400 cases per year in the United States. Less than one MCC is found for each 100 diagnosed melanomas [2].

It has very rarely been described on the fingers or the hand, with only few case reports that we are aware of [3–15,16]. Because of its rarity and the small numbers of patients in clinical reports, management is not standardized.

The tumor location has been categorized into three basic groups: head and neck, extremities, trunk.