Evolving paradigm in treatment of Merkel cell carcinoma of the digit from ray resection to wide local excision and flap coverage

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Abstract

Background Merkel cell carcinoma is a rare cutaneous neuroendocrine neoplasm that typically presents on the head and neck. Merkel cell carcinoma (MCC) of the digits is rare, and thus limited treatment options have been presented.

Methods In contrast to the current treatment recommendation of ray resection of the affected digit, two patients were treated by sentinel node biopsy, wide local excision, and flap coverage of the defect.

Results No local recurrences or metastases have been seen 24 months postoperatively.

Conclusion We suggest that the paradigm of ray amputation of a digit affected by MCC should evolve to initial treatment with a sentinel lymph node biopsy (SLNB) with subsequent local excision with wide margins and local flap coverage.

Keywords Merkel cell carcinoma · Digit · Hand · Treatment

Introduction

Merkel cell carcinoma (MCC) is an aggressive, neuroendocrine neoplasm first described by Toker in 1972 [1]. Over the past 15 years, the incidence of MCC has tripled [2, 3]. The

lesions are typically fast growing, non-tender, reddish-purple nodules found on the sun-exposed skin of Caucasian (95 %) males (61.2 %) over the age of 65 (80 %) [4]. A definitive diagnosis can be made using an immunohistochemical staining for cytokeratin 20 [2].

Even though the Surveillance, Epidemiology, and End Result (SEER) database reported 1234 (24 %) Merkel cell carcinomas in the hand and upper extremity from 1986 to 2009 [5], there are only 11 case reports of MCC of the digit found in PubMed, Medline, and MD Consult databases [6–16]. Although over a thousand cases were identified in the SEER database, there was no further subdivision of the upper extremity.

As outlined by Mikolyzk and Bednar in 2008 and confirmed by Parcells et al. in 2014, the current recommendation for treatment of MCC of a digit includes ray resection, sentinel lymph node biopsy (SLNB), and perioperative metastatic evaluation [10, 17]. This obviously removes the tumor, but at the cost of the digit. In contrast, our goal has been to preserve the finger as well as remove the invasive MCC. We believe that many cases of digital MCC can be treated by wide local excision, SLNB, and flap coverage.

Case 1

A 55-year-old woman presented with a 1-cm longitudinal scar on the dorsum of the proximal phalanx of her left middle finger six weeks after excision of a histologically proven MCC by a dermatologist. The patient had initially presented to the dermatologist with a non-tender bluish 10–12 mm lesion. Diagnosis of MCC was confirmed by immunohistochemical staining for cytokeratin 20. Six weeks after the excisional biopsy, a wide excision measuring 3×2 cm was performed down to the extensor tendon (Fig. 1). Sentinel lymph

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Fig. 1 Perioperative view showing excision to the extensor tendon before skin graft was harvested. Blue dye was used to find the sentinel lymph nodes

nodes were sampled and were negative for MCC. The defect was reconstructed using a reverse second metacarpal artery island flap (Fig. 2). Postoperatively, the flap healed uneventfully, and she regained full range of motion at the MCP, PIP, and DIP joints (Fig. 3). She remains disease free 26 months postoperatively.

Case 2

A 66-year-old woman presented after excisional biopsy of a 3×4 mm reddish lesion on the dorsum of the proximal phalanx of her right index finger by a dermatologist. The patient had a past medical history of a basal cell carcinoma of the skin, but the diagnosis of MCC was confirmed histologically by a high number of cytokeratin 20. Ten weeks after the excisional biopsy, a wide local excision of 2.5×2.5 cm was performed down to the extensor tendon. Sentinel lymph nodes were negative for MCC. The defect was reconstructed using a reverse second metacarpal artery island flap. Postoperatively, the flap healed primarily and she regained full

Fig. 2 Postoperative view showing intact reverse metacarpal artery flap

Fig. 3 Dorsal hand 3 months postoperative

range of motion of all joints in the affected finger. Metastatic work-up, including both a CT and PET scan, was negative. She remains disease free 24 months postoperatively.

Discussion

There is a significant discrepancy between the number of case reports of MCC in the digits and the number of MCCs reported in the SEER database for the entire upper extremity. Perhaps the majority of MCCs were located on the upper arm or forearm. There are 13 case reports of Merkel cell carcinoma localized to the fingers and 11 case reports of MCC on the dorsum of the hand. The purpose of this paper is to question the treatment of MCCs of the digit. Why remove a finger when it can be preserved? Is a ray amputation essential to prevent metastasis? One might argue that a ray resection is the safest procedure to remove a MCC and decrease the chance of metastasis. However, MCCs of the hand tend to be diagnosed at an earlier stage and consequently have lower rates of regional and systemic metastasis [4]. The true predictor of potential metastatic spread of MCC is the sentinel nodes. Although there is a positive correlation between the size of a MCC and the positivity of the SLNB, the results of the sentinel nodes, not the size of the lesion, should determine the treatment [18, 19].

Four previous case reports have documented that after a wide local excision of a digital Merkel cell tumor, a patient can remain tumor free. Brown et al. reported an intraepidermal MCC in the index finger, which was primarily excised and the function of the finger preserved [6]. Okada et al. reported a MCC in the small finger, which was excised and reconstructed with a stepladder dorsal metacarpal artery flap [11]. Spalveri et al. reported a 2-cm diameter MCC on the dorsum of the proximal phalanx of the index finger treated by wide local excision. The 3.5×3 cm defect was reconstructed with a retrograde kite flap, and the patient remained disease free 20 months postoperatively [15]. Su et al. reported a 4-mm lesion on the ring finger, which was excised, and the sentinel

nodes were negative. Six years later, there has been no recurrence [16].

Our two cases with both patients remaining disease free 2 years postoperatively as well as the previous four cases lend support to the argument that MCCs of the digit do not automatically mandate a ray amputation. We suggest that the paradigm of ray amputation should evolve to initial treatment with a SLNB despite the size of the MCC to check for metastatic spread. If the lymph nodes are negative, wide local excision of the MCC can be performed with immediate reconstruction with a local flap, such as a reverse metacarpal artery flap.

Conflict of Interest Alex Doermann declares that he has no conflict if interest.

James Jakowatz declares that he has no conflict if interest.

Neil Jones declares that he has no conflict if interest.

Statement of Human and Animal Rights All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent Informed consent was obtained from all patients for being included in the study.

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