Merkel Cell Carcinoma: A Special Case of Paronychia

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PURPOSE

Merkel Cell Carcinoma (MCC) a rare but aggressive type of skin cancer. The purpose of this case is to provide one instance of a unique case of MCC. While this condition is rare, the bleak prognosis merits a high level of suspicion from the onset.

LITERATURE REVIEW

MCC is a rare form of skin cancer that is very aggressive. It commonly affects Caucasian males between the ages of 70 and 80, only affecting 4% of people aging 49 and under (1). Discovery of the Merkel cell is attributed to Friedrich Merkel in the late 1800s, who described them as "touch cells." Today it is confirmed that Merkel cells relay sensory information corresponding to touch to the brain and are present in varying densities throughout the skin (2).

MCC is defined by the uncontrolled replication of infected Merkel cells located in the stratum basale of the epidermal skin. Typically appear on sun exposed areas, MCC presents as a "reddish glassy" appearance, with pale/violacious firm nodular dermal lesions (3). Lesions that are rapidly growing and riddled with telangiectasias, or are ulcerative can denote advanced stage of the disease (4). The eighth edition of the TNM staging system developed a staging system that also provides appropriate management guidelines. Stage I involves a primary lesion <2cm in size while Stage II describes a lesion >2cm with possible involvement of deeper tissues like bone, muscle, fascia, or cartilage. When there is primary lymph node involvement, that is considered as Stage III. Diffuse metastasis is Stage IV (4,5). This case represents a Stage II lesion without involvement of primary lymph node.

Increased association has been found in patients exposed to UV radiation. MCC by nature exhibits a predilection for sun exposed areas of the body. This characteristic is shared among numerous other malignant skin cancers. In a study by Engels et al where 309,365 patients with HIV were studied to find a 13.4% risk of MCC (6). In another study by Penn et al, it was shown that MCC was more aggressive in immune-compromised individuals, where 68% of the patients had lymph node metastasis and 56% died of the malignancy (7).

A variety of immunohistochemical and enzyme assays are used to detect expression of chromogranin, synaptophysin, and others to confirm MCC diagnosis. A variety of neoplasms display similar characteristics as MCC and as such diagnosing this ailment becomes very difficult. Cytokeratin (CK)20 positivity is specific to MCC and can be used as a diagnostic tool to distinguish MCC from other small round blue cell tumors (8).

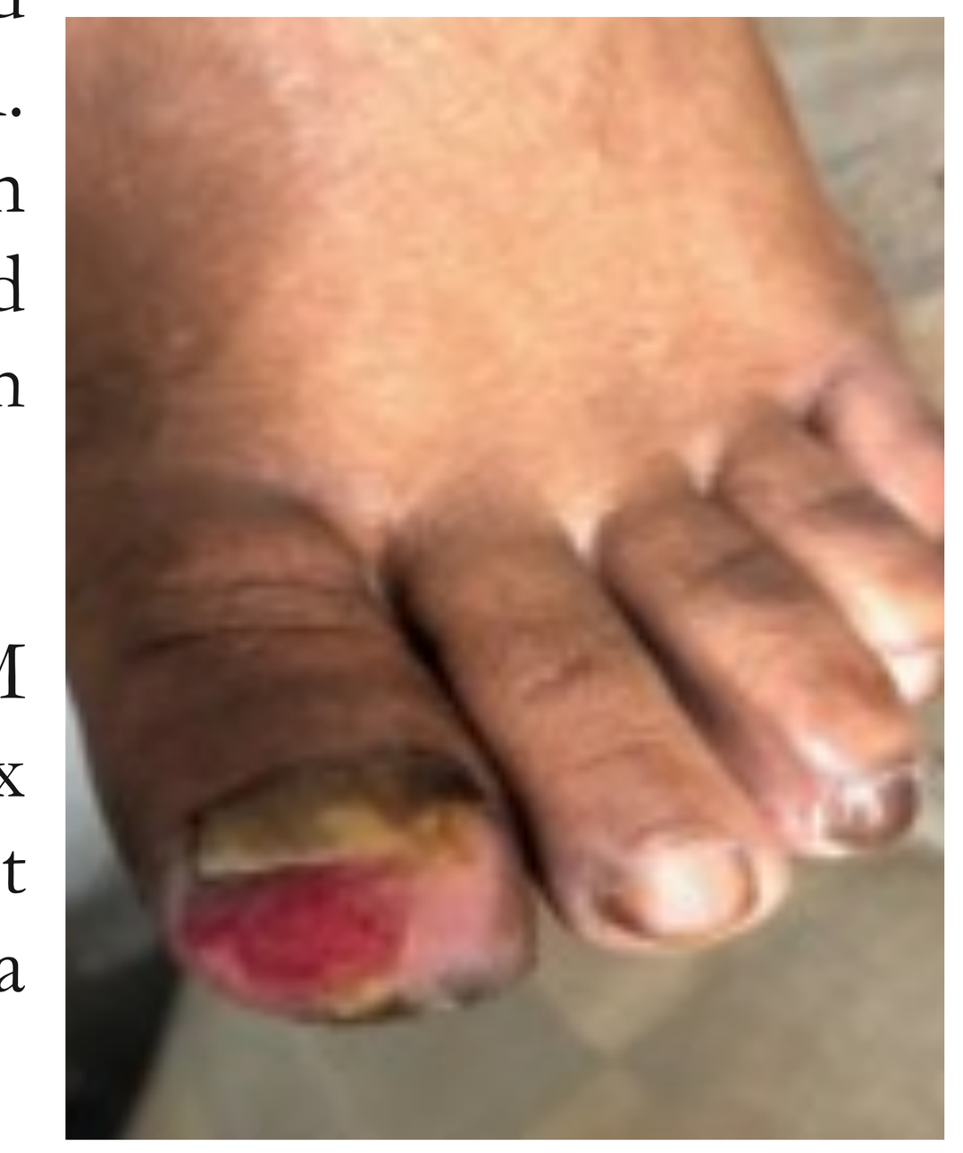
CASE STUDY & RESULTS

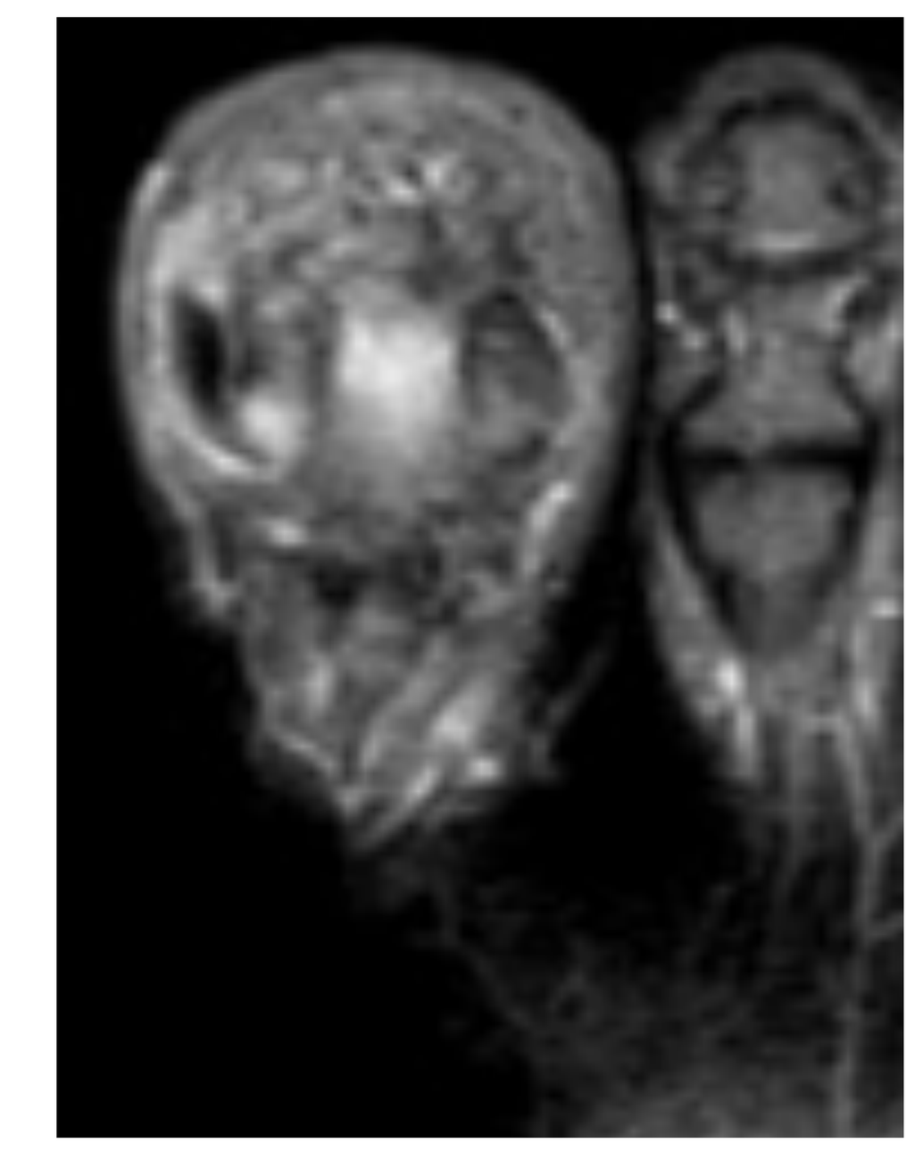
46-year-old female with Asthma, HIV seen with hallux nail bed chronic ulcer, possible osteomyelitis (OM) shown in Fig. 1A. She first ulcerated when she visited a nail salon, developed an infection. Over the next year, the ulcer would heal, but would then quickly re-open. On exam, a superficial ulcer, 2x2cm, with granular base, no probe to bone, no drainage, but very tender.

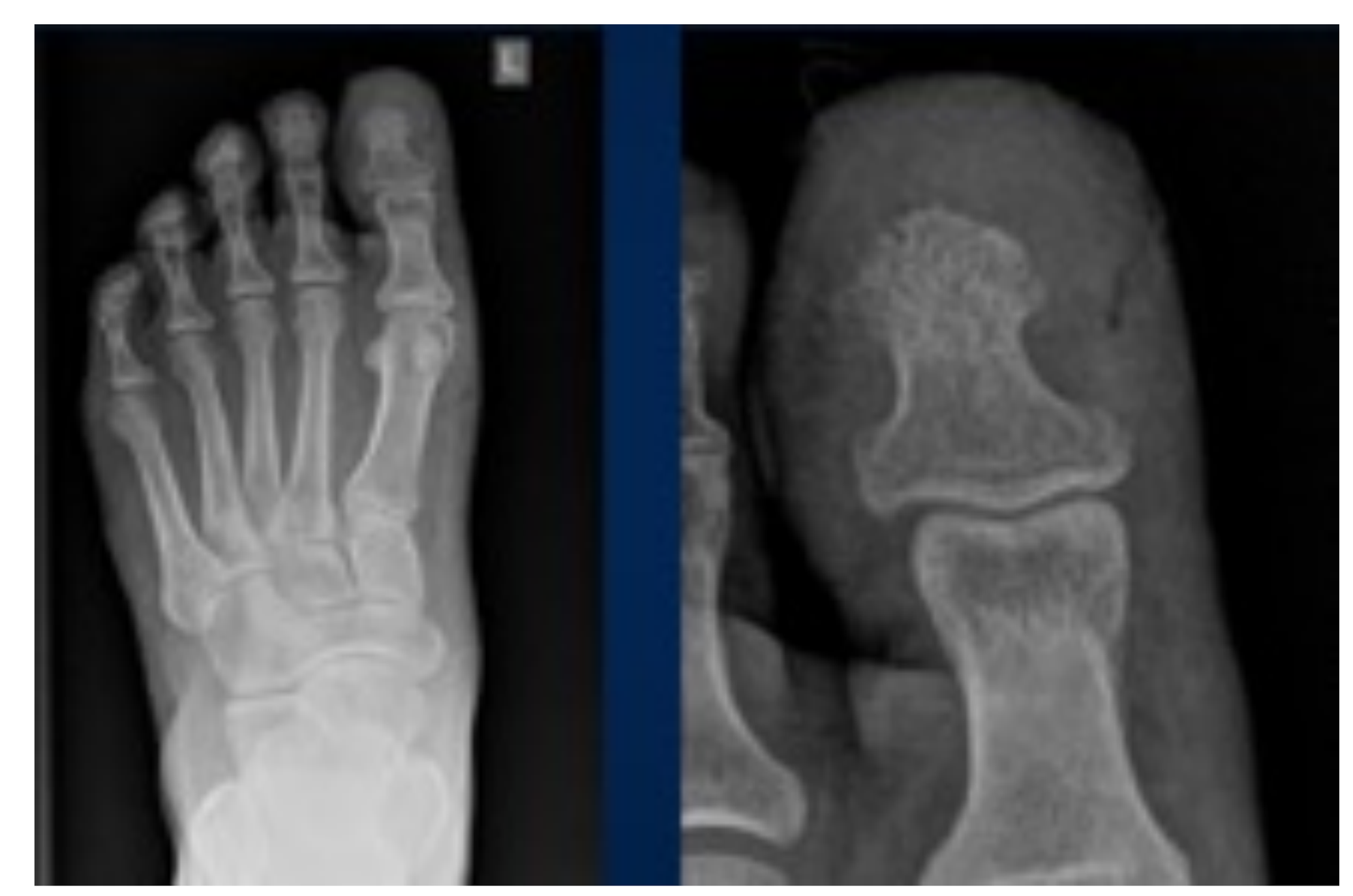
Radiographs showed no evidence of OM, MRI showed OM of the left hallucal distal phalanx. Distal tuft of distal phalanx was removed, a skin/bone biopsy performed. Pathology report confirmed this diagnosis of a neuroendocrine carcinoma specifically MCC.

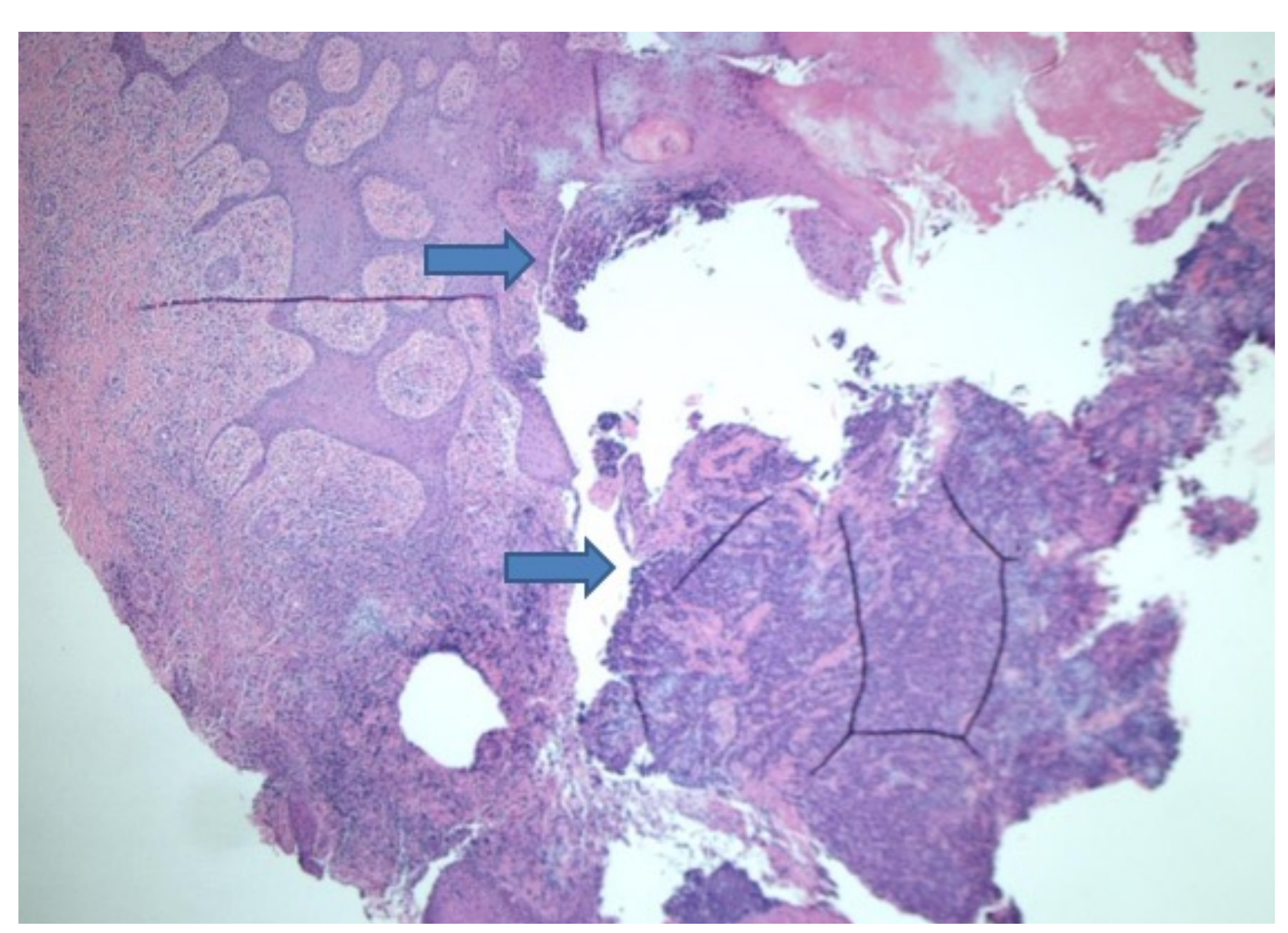
The skin biopsy showed neoplastic cells expressing neuron specific enolase, synaptophysin, and cytokeratins (AE1:3 and CK20) in a membranous and paranuclear, dot-like pattern. Chromogranin shows focal positive staining. The bone biopsy showed cytokeratin (AE1:3) stain highlighting the neoplastic cells. These studies were used as diagnostic tools to confirm MCC.

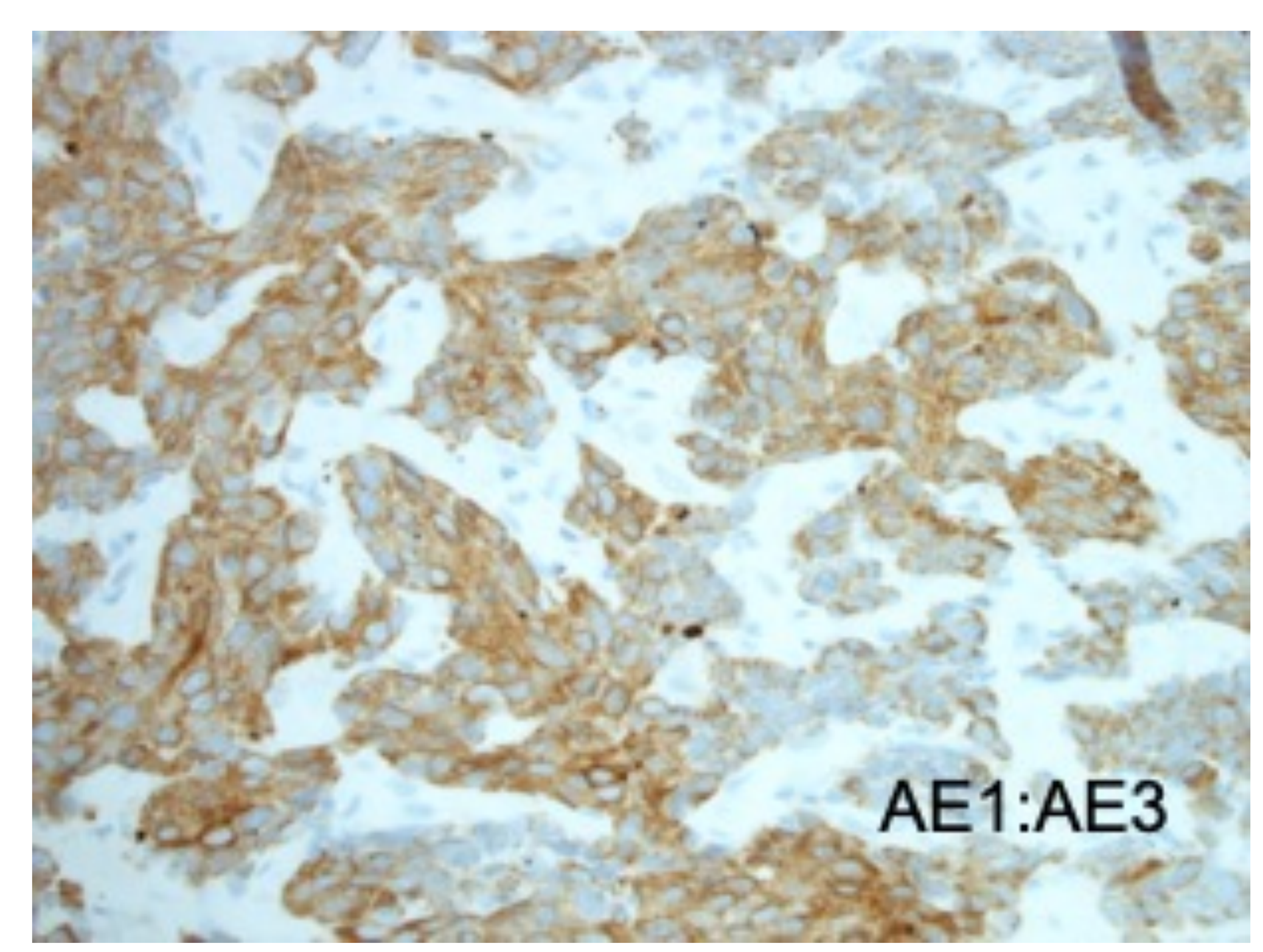
Patient was referred to an oncologist for continued care. Sentinel lymph node biopsy and a PET scan were negative for any metastasis. Patient underwent a hallux amputation with clean margins. Now doing well post operatively with no open lesions, minimal radiation therapy, and no chemotherapy.

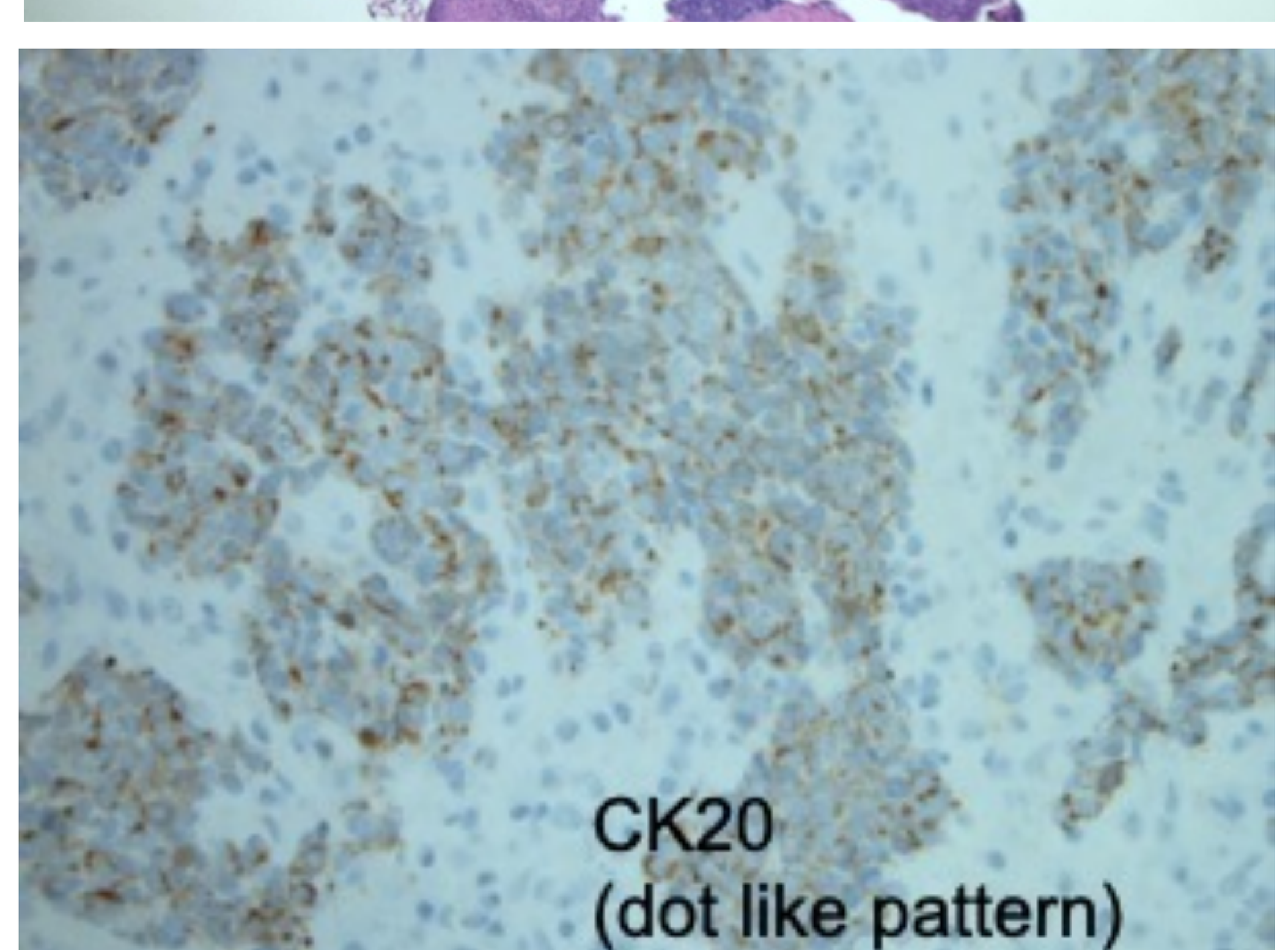


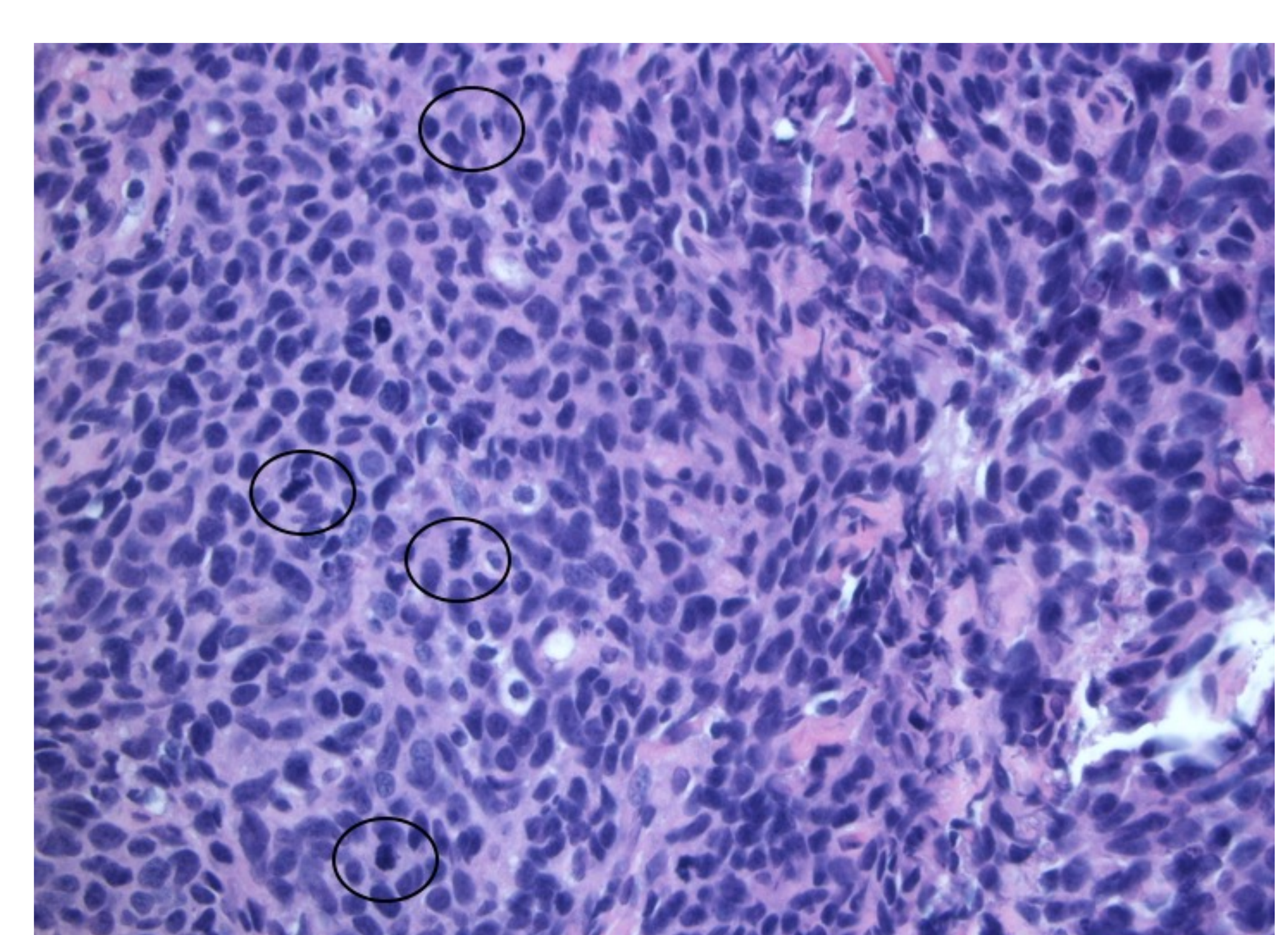












DISCUSSION

Goals of treating MCC include maximizing survival, minimizing recurrence, and improvement of morbidity and mortality associated with progression of this disease with metastasis. If diagnosed early while the tumor lesion remains localized and limited to cutaneous areas, patients have the same life-expectancy as those without the disease. Once MCC metastasizes to surrounding lymph nodes, the prognosis of the patient rapidly become bleak to a point where over 50% of patients succumb to disease progression to distant sites: bones, liver, lungs, brain, and distant lymph nodes (9,10).

Current treatment guidelines recommend wide 1-2cm local excision margins, and biopsy of sentinel lymph node irrespective of clinical presentation. Despite large excision margins, recurrence remains common and is attributed only partially to resection margins (9). In lymph node positive patients, lymph node dissection may be performed or radiotherapy to nodal basin (10).

MCC has shown to be high risk for metastasis, with the primary tumor size as the best predictor of risk. Even small tumor sized lesions are associated with 10-20% sentinel nodal metastasis. As such, detailed immunohistochemistry analysis of sentinel lymph nodes has now become imperative (10).

The patient in this case study presented with a very common ailment to a podiatrist, which is paronychia. The highly healthy and granular appearance of the lesion and its persistent nature for over a year were the two main clues to broaden the differential. It is suspicious that an ulcer with completely granular base, with normal underlying vascular supply, was persistently non-healing. The only characteristic marker for MCC this patient meets is being immuno-compromised secondary to HIV. In similar patients, lesional biopsies and efficient multidisciplinary involvement should be the standard of care.

It requires a high index of suspicion from the treating physician. Patients diagnosed in the early stages of the disease have a significantly more promising prognosis especially in comparison to those in later stages. AEIOU is an easy method to describe a MCC lesion: Asymptomatic lesion, Expanding rapidly, Immune suppression/compromise, Older age >50years, UV exposure history (11).

In this case, the high index of suspicion, prompt decision to biopsy the lesion, and quick involvement of the oncologist led to the early diagnosis of this malignant neoplasm and early treatment of the patient.

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