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Polydactylous Longitudinal Melanonychia Acquired following Total Skin Electron Beam Radiation Therapy for Sézary Syndrome

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To the Editor,

Longitudinal melanonychia (LM), the presence of longitudinally oriented pigmented band(s) along the nail unit, can occur in a single or multiple nail units, and is most commonly seen in individuals with skin phototypes 4–6 [1]. We would like to call to your attention the case of a 60-year-old Caucasian woman with no prior abnormalities of the nails, who presented with a 2-year history of progressive generalized erythema and keratoderma of her palms and soles. She had been seen by multiple physicians who diagnosed her as having a photosensitivity reaction. Discontinuation of her hydrochlorothiazide and treatment with low-dose prednisone did not result in any significant improvement. Subsequently, a skin biopsy revealed lymphocytic atypia with a skewed CD4:CD8 ratio, and PCR-based T-cell receptor gene rearrangement studies showed matching T-cell receptor clones in the skin and blood. Flow cytometry of her peripheral blood showed an aberrant CD3+/CD4+CD26– lymphocyte population (absolute count of 2,600 cells/ μ l). These findings supported a diagnosis of Sézary syndrome.

The patient was initially treated with bexarotene at a daily dose of ~ 200 mg/m² in conjunction with 100 μ g of interferon gamma 3 times a week, daily applications of mechlorethamine 0.01% in petrolatum, clobetasol 0.05% ointment for her palms and soles, and extracorporeal photopheresis for 2 consecutive days every 4 weeks. Six months after starting this regimen, the patient continued to have erythroderma and significant blood involvement on flow cytometry. Therefore, the decision to add total skin electron beam radiation therapy (TSEBT) to her regimen was made. The patient received 40 Gy worth of radiation to her body and an additional 10 Gy to her feet. During the 20 weeks of radiation treatment, the patient continued with her multimodality immunotherapy. One month after completing TSEBT, 85% of her skin had cleared and her Sézary count on flow cytometry dropped to 150 cells/ μ l. Secondary to the radiation therapy, the patient lost all of her scalp and body hair and all of her fingernails and toenails.

Two months after the completion of TSEBT, her nails started to grow back. Notably, a discrete 4-mm longitudinal brown streak was appreciated on the nail unit of her right second finger. Two

Fig. 1. Representative photographs of fingernails (a) and toenails (b) showing discrete longitudinal pigmented bands of varying widths. Asterisks represent involved nail units.

weeks later, examination of all 20 nails revealed longitudinal pigmentation in 13 of 20 nail units with bands of different widths ranging from 2 to 10 mm. There was no pigmentation on the nail folds (fig. 1). Given the polydactylous multiple nail involvement and the recent onset of these findings, we recommended baseline photographs and close monthly monitoring. We find this unique observation of acquired polydactylous LM as a side effect of TSEBT important for physicians to understand. There is a single report of 3 cases of transverse melanonychia on multiple fingernails following TSEBT for mycosis fungoides (1 African-American, 2 Caucasian) where the pigmentation was displaced outwards as the nails grew [2]. However, this is the first case of LM seen after TSEBT for cutaneous T-cell lymphoma (CTCL).

Nail dystrophy and temporary anonychia are common side effects of TSEBT occurring in ~40 and 16% of treated patients, respectively [3, 4]. LM in the setting of radiation therapy could be the result of various elements affecting the distribution or production of melanin. The rapid development of LM is suggestive of postinflammatory pigment alteration of the nail unit secondary to TSEBT as it is frequently seen in other areas of the skin in postradiated CTCL patients. We hypothesize that electrons can induce local inflammation, which can contribute to subsequent release of pigment. Alternatively, electrons could be altering the transcriptional activity of previously dormant melanocytes or inducing posttranslational modifications resulting in increased numbers of melanosomes.

As mentioned above, in addition to TSEBT, this patient had also been treated with the retinoid X receptor agonist bexarotene and superpotent topical steroids for several months. Adrenal insufficiency, a known side effect of both aforementioned treatments, has been linked to the development of LM [3]; in this case, the patient had normal nails until she underwent TSEBT. Furthermore, her a.m. cortisol levels had been checked periodically and found to be within normal limits. While reports of melanonychia associated with photochemotherapy have been published [4, 5], it is unlikely that the negligible concentration of psoralen in the blood after extracorporeal photopheresis treatments may have played a role in the development of polydactilous melanonychia.

In summary, this first report of LM following TSEBT in a Sézary patient should alert the general dermatologist and CTCL

experts to carefully examine each of the nail units prior to TSEBT. This observation could provide the basis for a larger longitudinal study to follow patients after TSEBT in order to evaluate the frequency of LM as a potential complication in the short or long term.

Statement of Ethics

The patient was seen in our practice as part of routine care, she gave informed consent for use of the photograph, and her human rights were respected.

Disclosure Statement

None of the authors has any potential financial conflict of interest related to this paper.

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