Widespread Keratotic Spiky Follicular Papules Associated With Hyperpigmentation: Challenge

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CASE REPORT

An 8-year-old boy, who has been receiving chemotherapy for T-lymphoblastic lymphoma (T-LBL) for the last 3 years, presented with a 2-year-history of a widespread skin eruption. The condition started 1 year after the onset of chemotherapy when itchy lesions appeared over his face, described by his parents as "pimple like." They progressed gradually and within 3 months, similar lesions appeared over both upper limbs, lower limbs, and trunk. On examination, follicular papules with keratotic/spiky plugs were distributed over the face, mainly over the nose, nasal vestibule, malar area, forehead, eyelids, eyebrows, nape, and ears (Figs. 1A–C) with alopecia affecting the eyebrows and eyelashes. The trunk, thighs, legs, feet, arms, forearms, and hands were involved by similar lesions but to a lesser extent, and lesions on the trunk and extremities were associated with a background of hyperpigmentation (Fig. 1D). Scalp hair was normal. The patient found the lesions cosmetically unsightly and was used to plucking out the spikes.

Three punch biopsies were taken, one from a keratotic spiky papule on the nape and 2 from keratotic spiky papules with hyperpigmented background on the left thigh and trunk. All biopsies showed abnormally looking dilated hair follicles with expanded inner root-sheath cells having characteristic large eosinophilic trichohyalin granules replacing follicular lumina with absent or poorly formed hair shafts (Fig. 2). Associated epidermal changes of orthokeratosis, mild papillomatosis, and mild epidermal hyperplasia were also detected. In addition, biopsies taken from lesions with background pig-

FIGURE 1. Keratotic spiky papules over the face (A). Ears are also involved (B, C). Lesions on the trunk and extremities are associated with background pigmentation (D).

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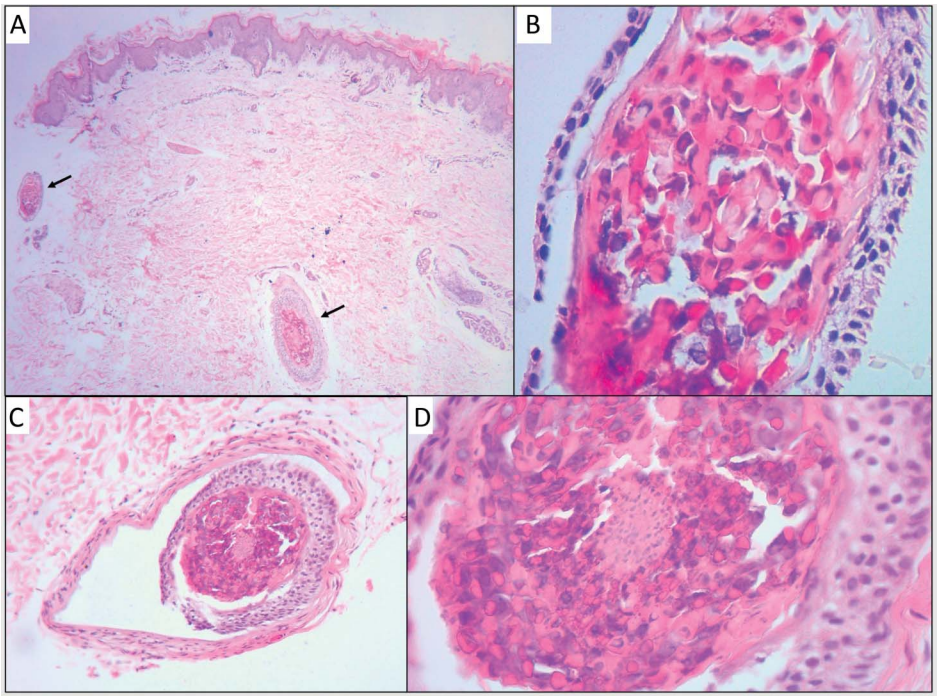

FIGURE 2. A, B, Histopathological features of a biopsy taken from one of the keratotic lesions showing abnormally looking dilated hair follicles with expanded inner root sheath (black arrows). Higher magnification shows large eosinophilic trichohyalin granules replacing the follicular lumen with absent hair shaft (A: H&E ×40, B: H&E ×400). C, D, Another biopsy showing similar features with a poorly formed hair shaft (C: H&E ×100, D: H&E ×400).

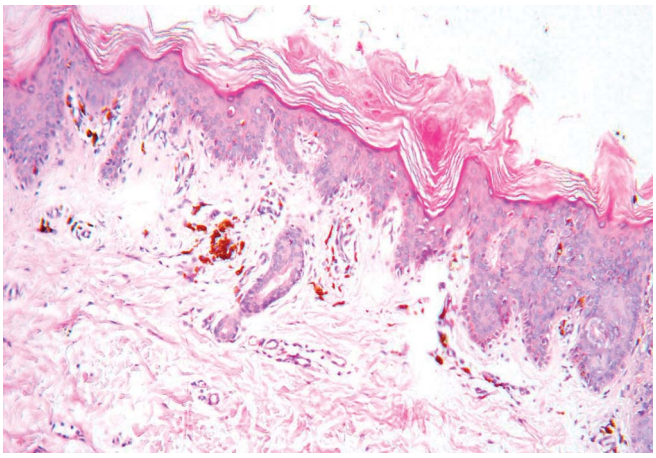

FIGURE 3. A biopsy taken from a keratotic lesion with background of pigmentation showing scattered necrotic keratinocytes within the epidermis and stratum corneum in addition to many melanophages (H&E ×100).

mentation showed many necrotic keratinocytes within the epidermis, and a superficial perivascular lymphohistiocytic infiltrate admixed with many melanophages (Fig. 3).

WHAT IS YOUR DIAGNOSIS?

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