Case of the Month

Diagnosis: Spiny Keratoderma

S T O W E , V T. — The differential diagnosis for this patient included punctate porokeratosis, verruca vulgaris, pitted keratolysis, arsenical keratoses, and genetic disorders such as Darier’s disease and Cowden disease that can cause keratotic lesions, Dr. Jamie A. Alpert said at a dermatology conference sponsored by the University of Vermont.

Histologic features of her lesions included a discrete, compact parakeratotic column with slight epidermal invagination that was contiguous with the granular layer and easily differentiated from the adjacent orthokeratotic layer. The epidermis was otherwise unremarkable, and there was no appreciable inflammation.

The diagnosis was spiny keratoderma, an autosomal dominant condition characterized by numerous tiny keratotic projections that resemble the spines on the rotating drum inside a music box. The disorder can be palmoplantar or generalized and is associated with malignancy in a subset of patients, Dr. Alpert of the University of Vermont, Burlington, said.

Many reports of palmoplantar punctate keratotic projections have appeared in the literature, dating back to 1971. In addition to the name spiny keratoderma, these lesions also have been called punctate keratoderma, punctate porokeratosis, palmoplantar keratosis acuminatum, and palmoplantar filiform hyperkeratosis.

Broad classification of palmoplantar punctate keratotic lesions is based on morphology, distribution, and whether they are acquired or inherited.

In 1994, Dr. Thomas W. McGovern devised a classification scheme specifically for spiny keratoderma (SK), which he named “music box spine dermatoses.” He divided the various presentations into five groups, based on location and histology:

1a: Palmoplantar parakeratotic SK, involving predominantly the palms and/or soles, with histology showing the parakeratotic column. The patient had this type.

1b: Disseminated parakeratotic SK, which was widespread with palmoplantar sparing, with the same parakeratotic column as is seen in the palmoplantar parakeratotic type.

2a: Palmoplantar orthokeratotic SK, involving predominantly the palms and/or soles and showing orthokeratotic hyperkeratosis on histology.

2b: Disseminated orthokeratotic SK, widespread with general palmoplantar sparing and with histology showing orthokeratotic hyperkeratosis.

3: SK in eccrine hamartoma, which can occur on any cutaneous surface and shows a column of parakeratosis associated with skin appendages on histology.

Treatment is difficult. Lanolin and petrolatum-containing moisturizer may provide some relief. A variety of other agents have been tried, including retinoids, tazarotene, salicylic acid peel, fluorouracil cream, and 6% salicylic acid (usually under occlusion), as well as electrodesiccation and mechanical removal.

“They all work to some degree, but it always comes back,” Dr. Alpert said.

An age-appropriate malignancy workup should be considered in all patients with new onset of SK and widely distributed lesions. This patient underwent a complete blood count with differential, metabolic panel, chest x-ray, mammogram, and colonoscopy. No internal cancers were identified.

Treatment with 40% urea cream smoothed out her lesions, and she has not been seen since February 2006.

She should return with recurrence, the plan is to try tazarotene gel or use a Dremel tool to sand down the lesions, Dr. Alpert said in an interview.

—Miriam E. Tucker

TREATMENT

For topical use only, not for intracutaneous or oral use. (See INDICATIONS AND USAGE.)

FOR TYPHOID ONE OR THE OTHER, NOT FOR BOTH.

BRIEF SUMMARY

RENOVA (tretinoin cream) 0.02%

INDICATIONS AND USAGE

RENOVA cream 0.02% is indicated as an adjunctive therapy for the treatment of acne vulgaris. RELEVANT PHARMACODYNAMIC AND PHARMACOKINETIC DATA

When using RENOVA cream, patients should take steps to prevent phototoxicity. Use sunscreen, protective clothing, protective gloves, and occlusion wherever possible.

PRECAUTIONS

General

RENOVA should be used on an as-needed basis to treat only the affected skin areas. Avoid the eyes and mucous membranes, as tretinoin may cause irritation.

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C A S E O F T H E M O N T H

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COURTESY DR. D E B O R A H L. C O O K

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