Linear Porokeratosis

Excellent Response to Photodynamic Therapy

Xavier García-Navarro, MD; Joan Ramon Garcés, MD; Eulàlia Baselga, MD; Agustín Alomar, MD; Department of Dermatology, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain

Linear porokeratosis is a skin condition that usually shows a poor response to different modalities of treatment. We describe a patient successfully treated with 2 sessions of photodynamic therapy (PDT).

REPORT OF A CASE

A 13-year-old boy presented with a 4-year history of reddish-brown hyperkeratotic papules and plaques on the back of his lower leg. The lesions were arranged in a linear pattern and had a raised peripheral ridge and atrophic center (Figure 1). There was no nail dystrophy or other associated abnormalities. His mother had disseminated superficial actinic porokeratosis (DSAP) in the lower extremities. The patient's skin lesions were asymptomatic but cosmetically distressful. Findings from a histopathologic examination showed epidermal hyperplasia with a vertical column of tightly packed parakeratotic cells, the cornoid lamella. These findings were consistent with linear porokeratosis. No response was observed after 1 year of treatment with calcipotriol ointment.

THERAPEUTIC CHALLENGE

There is no known cure for linear porokeratosis, and treatment generally produces disappointing results. However, multiple topical therapies, such as keratolytics, corticosteroids, retinoids, fluorouracil cream, and imiquimod or calcipotriol cream, have been used with variable results. Oral acitretin has proven effective in widespread disease, but recurrence following discontinuation is common. Other modalities, such as Q-switched ruby laser, fractional photothermolysis, cryotherapy, carbon dioxide laser ablation, curettage, or dermabrasion, can be useful in localized lesions but are not feasible for larger lesions owing to the risk of scarring.

Figure 1. Reddish-brown hyperkeratotic papules and plaques linearly arranged on the back of the right lower leg before treatment.

Figure 2. Erythema and inflammation within the lesions after the first session of photodynamic therapy and methylaminolevulinate hydrochloride cream.

Figure 3. Clinical aspect of the treated zone 11 months after the second session of photodynamic therapy and methylaminolevulinate hydrochloride cream.
After removing the superficial scales with a carbon steel surgical blade, methyl aminolevulinate hydrochloride cream in a 160 mg/g concentration (Metrivix; Penn Pharmaceuticals Services, Gwent, South Wales) was applied under occlusion in the medial part of the lesion for 3 hours. Subsequent illumination with red light (light dose of 37 J/cm²) (Aktilite; Photocure ASA, Oslo, Norway) was performed for 9 minutes (Figure 2). Irradiation was well tolerated, and anesthesia was not necessary because the patient noticed only a mild burning sensation. A second session was performed 1 month later. The cosmetic and clinical response was excellent, and the patient was highly satisfied. No recurrence was observed after 11 months (Figure 3), and we are presently treating the remaining lesion.

Porokeratosis is a group of genetic disorders characterized by clonal proliferation of keratinocytes. Five variants have been identified to date. These include porokeratosis of Mibelli, DSAP, punctate porokeratosis, porokeratosis palmaris et plantaris disseminata, and linear porokeratosis. Linear porokeratosis is a rare clinical variant that usually arises in childhood. It commonly appears on the extremities as unilateral, linear hyperkeratotic papules and annular plaques. Lesions may coalesce into larger plaques with central atrophy and a raised border along the lines of Blaschko. Nail dystrophy has also been associated with this disorder. Linear porokeratosis seems to have an increased risk of transformation into squamous cell carcinoma or basal cell carcinoma and may be exacerbated by immunosuppression, UV light, or irradiation. Differential diagnosis includes linear verrucous epidermal nevus, lichen striatus, incontinentia pigmenti, linear lichen planus, linear Darier disease, and warts.

Photodynamic therapy uses light to activate a photosensitizer in diseased tissue, leading to the formation of cytotoxic reactive oxygen species and selective cell damage. At present, it is primarily used to treat actinic keratosis and superficial nonmelanoma cancers. We found only 2 reports about treatment of DSAP with PDT. Both were in adults, and results were contradictory. In the report by Nayeemuddin et al., no response was observed in 3 patients with clinical DSAP after treatment with PDT and 5-aminolevulinic acid. Cavicchini and Tourlaki, however, obtained an excellent clinical and cosmetic outcome in a patient with DSAP treated with PDT and methyl aminolevulinate cream. These different results may be related to the higher lipophilicity profile of methyl aminolevulinate, a drug that may perhaps penetrate hyperkeratotic lesions of porokeratosis more easily than 5-aminolevulinic acid hydrochloride. Treatment with PDT and methyl aminolevulinate cream was administered to our patient because this modality is routinely used for other indications in our setting and because of the efficacy reported by Cavicchini and Tourlaki in another type of porokeratosis.

To our knowledge, this is the first report of linear porokeratosis successfully treated with PDT. In our opinion, PDT together with methyl aminolevulinate cream could be an effective and safe alternative to conventional treatments for this disorder. However, controlled studies with a greater number of patients and long-term follow-up are needed to evaluate its real effectiveness. In our patient, no lesion recurrence was observed 11 months after treatment. Further follow-up is needed to determine whether additional sessions are needed to maintain the clinical response.

Accepted for Publication: June 5, 2008.
Correspondence: Xavier García-Navarro, MD, Department of Dermatology, Hospital de la Santa Creu i Sant Pau, Sant Antoni M. Claret 167, 08025 Barcelona, Spain (xgarcian@gmail.com).

Author Contributions: Dr García-Navarro had full access to all data in the study and takes responsibility for its integrity and the accuracy of the data analysis. Study concept and design: García-Navarro, Garcés, Baselga, and Alomar. Acquisition of data: García-Navarro. Analysis and interpretation of data: García-Navarro. Drafting of the manuscript: García-Navarro and Baselga. Critical revision of the manuscript for important intellectual content: Garcés and Alomar. Administrative, technical, and material support: García-Navarro. Study supervision: Garcés and Alomar.

Financial Disclosure: None reported.

REFERENCES

©2009 American Medical Association. All rights reserved.