CASE 19

Patient: A 18-year-old Thai female

Chief Complaint: Pruritic skin lesions for 1 year

Present Illness: The patient presented with erythematous pruritic papules confluent to plaques on her left arm and gradually formed linear brownish keratotic plaque on her left side of trunk, back and left thigh. The lesion was intensely pruritic and various medications were previously tried without clinical improvement. There was no history of vesicular eruption or association by sunlight.

Past History: Asthma

Family History: Unremarkable

Dermatological Examination (Figure 19.1-2)**:** Multiple light brown to erythematous papules along Blaschko's lines on Lt arm and confluent to sharply demarcated plaques with raised keratotic edge on left side of trunk

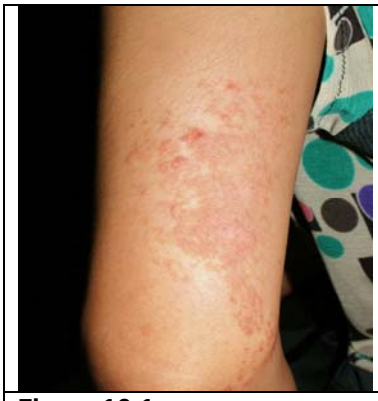

Figure 19.1

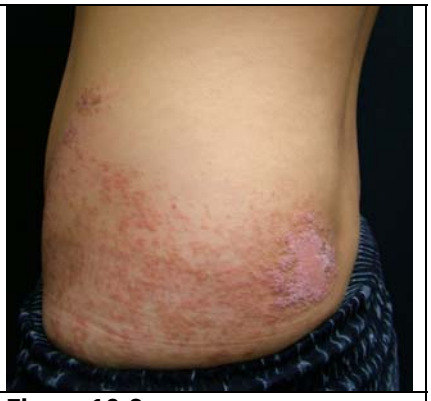

Figure 19.2

Physical Examination: Unremarkable

Histopathology (S09- 13086) (Figure 19.3): Tall columns of parakeratosis (cornoid lamellae) with hypogranulosis, dyskeratosis and vacuolated cells in the underlying atrophic epidermis

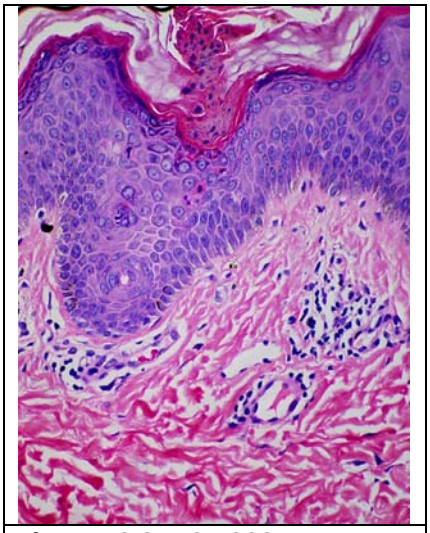

Figure 19.3, H&E 200x

Diagnosis: Linear Porokeratosis

Treatment: 0.05% Clobetasol propionate cream twice daily
Topical Calcipotriol cream twice daily
Topical 5% Imiquimod 5 times/week
CO_2 laser
Antihistamine
Closed monitoring for skin cancer

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Discussion:

The porokeratoses are a group of disorders of keratinization characterized clinically by sharply demarcated, hyperkeratotic, annular lesions with distinct thread-like keratotic edge corresponding histologically to the presence of cornoid lamella, a column of parakeratotic cells extending through the stratum corneum.[1]

Five clinical variants are recognized: (1).classic porokeratosis of Mibelli (2).disseminated superficial actinic porokeratosis and disseminated superficial porokeratosis (3).porokeratosis Palmaris et plantaris disseminata (4).punctate porokeratosis and (5).linear porokeratosis.[2] The etiopathogenesis is still unknown but certainly multifactorial. Porokeratosis is regarded as an autosomal dominant disorder with reduced penetrance except in linear type. There are many additional factors such as UV exposure, immunosuppression, and viral infection.

Linear porokeratosis is a rare variant that presents at birth or during childhood. The mode of inheritance has been suggested to be autosomal dominant,[3] but linear porokeratosis mostly occurs sporadically and might reflect mosaicism for a gene responsible for porokeratosis of Mibelli.

Two forms of linear porokeratosis exist. In the more common localized form of the disease, lesions are confined to an extremity, frequently distal and present unilaterally. In the rare generalized form, lesions are multiple, affect several extremities, and involve the trunk along Blaschko's lines.

Differential diagnosis of linear porokeratosis includes linear verrucous epidermal nevus, lichen striatus, incontinentia pigmenti, linear lichen planus, linear Darier disease.

There is no known curative therapy for linear porokeartosis. Treatment generally leads to disappointing results. Intervention is usually unnecessary, tumor transformation prevention by avoiding the former precipitating factors and tumor surveillance is standard. However, if the lesions are problematic or cosmetically unacceptable, multiple topical therapies such as keratolytics, corticosteroids, retinoids, fluorouracil cream, imiquimod and calcipotriol cream have been used with variable results.[4-6]

Photodynamic therapy together with methyl aminolevulinate cream could be an effective and safe alternative to conventional treatments for this disorder[7]. Other modalities, such as Q-switched ruby laser, fractional photothermolysis, cryotherapy, carbon dioxide laser ablation, curettage, or dermabrasion[5] can be useful in localized lesions but are not feasible for larger lesions due to the risk of scarring. Topical imiquimod cream could be considered as the promising therapeutic option in treatment of unresectable linear porokeratosis.[8]

Malignant degeneration has been observed in all five clinical variants of porokeratosis.[9] This includes Bowen's disease, squamous cell carcinoma, and rarely, basal cell carcinoma. Linear porokeratosis seems to be the most type of porokeratosis that can transform into squamous cell carcinoma or basal cell carcinoma. The transformation may be exacerbated by immunosuppression, UV light, or radiation.[10] The development of a nodule or a sore within a porokeratosis lesion warrants biopsy evaluation. The high risk factors are large, long-standing, or linear lesions.[11] Closed monitoring of malignant degeneration is very important in this patient.

References

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