CASE 1

A 13 years-old Thai female from Sakeaw

Chief complaint:

Generalized hyperkeratotic lesions for 11 years

Present illness:

11 years ago, when she was 2 years old, she has first developed hypopigmented annular patches at her left fingers and gradually formed linear brownish keratotic plaque on her left upper extremity, then the lesion spread progressively on her face, trunk, both upper and lower extremities. She occasionally feel itchy but she has no history of vesicular eruption or association by sunlight.

Past history:

Unremarkable

Family history:

Negative

Physical examination:

Skin: Generalized sharply demarcated, brownish, linear, hyperkeratotic plaque along Blaschko's lines with raised, thread-like border, and hypopigmented atrophic center at face, trunk, upper and lower extremities

All nails, teeth, hair are normal.

Histopathology: (S05-05548)

- Tall, thin column of parakeratosis
- Delling epidermis with dyskeratotic cells beneath the parakeratotic column

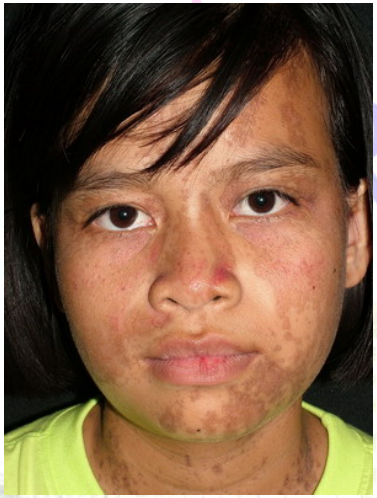

Fig. 1.1

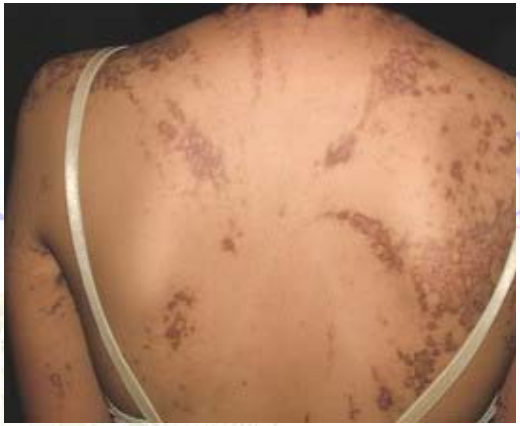

Fig. 1.2

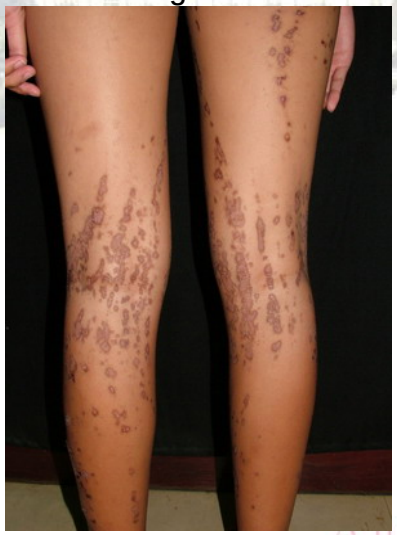

Fig. 1.3

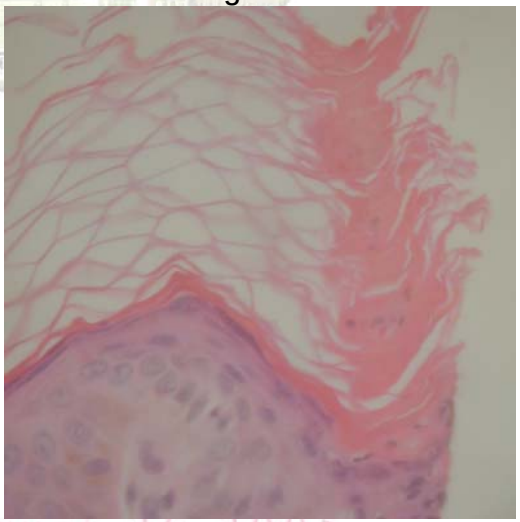

Fig. 1.4

Diagnosis :

Porokeratosis (Generalized linear porokeratosis)

Treatment :

Topical calcipotiol twice daily.

Closed monitoring for skin cancer.

Presesenter: Suthinee Kunkonkarn

Consultant: Natta Rajatanavin

Discussion:

The porokeratoses are a group of disorders of keratinization that has characteristic feature in thread-like raised hyperkeratotic border and presence of cornoid lamella in histology.

Five clinical variants are recognized: classic porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, linear porokeratosis, porokeratosis palmaris et plantariset disseminata, and punctata porokeratosis.

The etiopathogenesis is still unknown but certainly multifactorial. Porokeratosis is regarded as an AD disorder with reduced penetrance except in linear type. There are many additional factors such as UV exposure, immunosuppression, and viral infection.

Linear porokeratosis[1] is a rare distinctive variant of porokeratosis that presents at birth or in childhood as a linear verrucous eruption that may follow Blaschko's lines. Two forms of linear porokeratosis exist. In the more common localized form of the disease, lesions are unilateral, confined to an extremity, and frequently distal. In the rare generalized form, lesions are multiple, affect several extremities, and involve the trunk along Blaschko's lines like in this patient.The clinical differential diagnosis includes linear annular lichen planus, linear verrucous epidermal nevus, and lichen striatus.

Therapy is disappointing for all forms of porokeratosis. Some medications have been reported of potential benefit such as topical 5-fluorouracil, topical vitamin D_3, topical imiquimod, topical[2] and oral retinoid[3]. Several reported surgical modalities are excision, cryotherapy, electrodesiccation and curettage, diamond fraise dermabration, Grenz ray radiation and laser therapy such as CO_2 laser, 585nm flashlamp pumped pulse dye laser, and 532nm Nd:YAG laser.

Malignant degeneration[4] has been observed in all five clinical variants of porokeratosis. This includes Bowen's disease, squamous cell carcinoma, and, rarely, basal cell carcinoma. The high risk factors are large, long-standing, or linear lesions[5]. Closed monitoring of malignant degeneration is very important in this patient.

References:

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4. Sasson M, et al. Porokeratosis and cutaneous malignancy. Dermatol Surg 1996;22:339-342.
5. Curnow P, Foley P ,Baker C. Multiple squamous cell carcinomas complicating linear porokeratosis. Austral J Dermatol 2003 ;44:136–139.