CASE 5

A 30-year-old Thai female accountant from Bangkok

Chief complaint:

Hyperpigmented patches at legs and arms for 4 years

Present illness:

She had symmetric brownish reticulated hyper-pigmented patches at both legs and arms for 4 years. It first appeared on the lower extremities with mildly itching.

She had no history of atopy, no history of excessive sun exposure, no arthralgia, no fever, and no Raynaud's phenomenon. No history of prolonged rubbing of the extremities.

Past history:

No underlying disease

Family history:

none

Physical examination:

Skin: symmetrical reticulated brownish hyperpigmented macule with ripple-like pattern confluent to patches with few hypopigmented macules ontop at flexor and extensor surface of both lower and upper extremities.

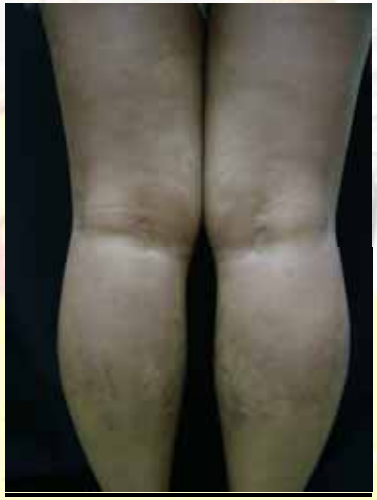

Fig. 5.1

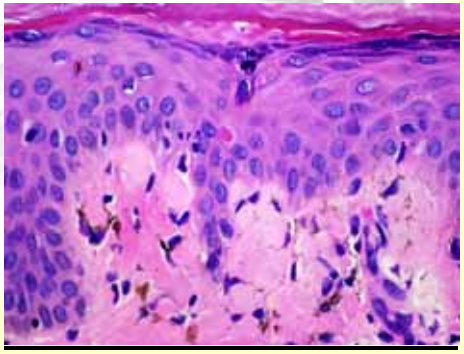

Fig. 5.2

Lab investigation:

- CBC Hb 13.4 g/dl ,Hct 39%,WBC 8170 /mm3,PMN 64%,Lymphocyte 28%,monocyte 6%, eosinophil 2%, Platelet 293,000
- Urinary examination WNL
- Coagulogram :WNL,
- ANA Negative, Anti DNA negative,Anti-HCV negative,HBsAg negative
- LFT:WNL, BUN 9 mg/dl,Cr 0.8 mg/dl

Histopathology: (S06-08525)

Small deposit of amphophilic globule in the broadened dermal papillae. Melanophage and stellate fibroblast observed within the globule.

Diagnosis: Amyloidosis cutis dyschromica with reticulate lesion
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Treatment: Topical triamcinolone acetonide 1% and 10% urea cream

Discussion:

Primary cutaneous amyloidosis is a rare,chronic,progressive skin disease primarily affecting adults. The name refer to deposition of amyloid in previously normal skin with no evidence of systemic involvement. Classically, macular, lichen and nodular form are the main types; poikiloderma-like, bullous, vitiliginous and anosacral forms are extremely rare clnical variants.

Amyloidosis cutis dyschromica defined by Morishima is assumed to be a specific type of vitiliginous amyloidosis. Clinically, it is characterized by a dotted dyschromia with reticular hyper- and hypopigmentation, mild or no itching, prepubertal onset and minimally deposited amyloid material just beneath the epidermis.

Genetic factor, hypersensitivity to UVB radiation, and possible DNA repair defects have all been incriminated as etiological factors.

The diagnosis of cutaneous amyloidosis depends on the histochemical, immunohistochemical, or ultrastructural demonstration of amyloid in the skin biopsy specimen.

Amyloid deposits usually are confined to the papillary dermis and do not involve blood vessels or adnexal structure. Early lesions contain small, multifaceted, amorphous globules within the papillae. These are missed easily without the use of special stain such as Congo red, Crystal violet, triphenylmethane dyes and thioflavine T under fluorescent light microscope. Later lesions show globules that coalesce, expand the papillae and displace the rete ridges laterally.

Various therapeutic modalities have been used in the treatment of cutaneous amyloidosis with variable success. For relief of symptoms; topical corticosteroids, keratolytic, dimethyl sulfoxide, capsaicin, carbondioxide laser, etretinate and acitretin have all been tried with variable results

References:

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5. Ozcan A, Mustafa S, Aydin NE, Karaca S. Amyloidosis cutis dyschromica: A case treated with acitretin,J Dermatol 2005,32:474-7