CASE 6

A 33-year-old Thai woman, from Khon-Kaen

Present history:

The patient presented with multiple discrete hyperpigmented and hypopigmented macules mainly on the extremities and back for 20 years. The lesions have gradually increased in the number and sometimes associated with mild pruritus.

Past history:

She is otherwise healthy.

Family history:

No family history of similar skin lesion.

Physical examination:

Vital signs: normal
HEENT: not pale, no icteric sclerae
Heart & lung: WNL
Skin: Multiple discrete sharp demarcated hyperpigmented and hypopigmented macules on both arms, both lower legs and back.

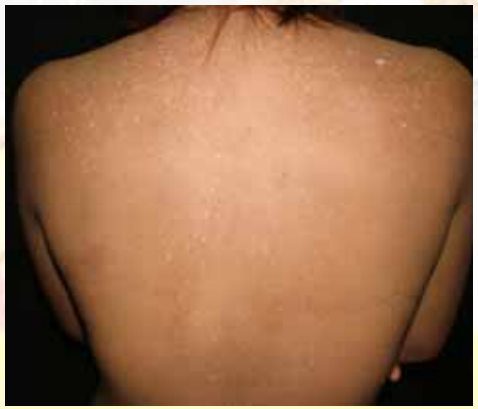

Fig. 6.1

Histopathology : (Slide No. S06-04721)

Small deposit of amphophilic globules with melanophages in the broadened dermal papillae.

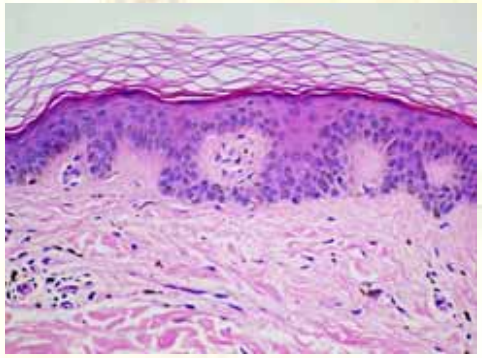

Fig. 6.2

Diagnosis: Amyloidosis cutis dyschromica
Presenter: Wiboon Thaneepakorn
Consultant: Siripen Puavilai

Discussion:

Amyloidosis cutis dyschromica first described by Morishma in 1970, is an uncommon variant of primary localized cutaneous amyloidosis. It is characterized by generalized dotted and reticulated macules of hyper-pigmentation with hypopigmented spots primarily in sun exposed skin with no or slight itching, usually onset before puberty. Small foci of amyloid deposits in the papillary dermis, and fibroblasts show hypersensitivity to UVB.

Although the exact etiology of amyloidosis cutis dyschromica is still unknown, it is assumed to be a congenital disorder with sun exposure as a major causative factor. This familial disorder has been reported mostly from Japan. Genetic factors lead to ultraviolet sensitivity and DNA repair defects. This repeated damage to the keratinocytes results in the promotion of amyloid materials in the skin. Amyloid deposits may initially be derived from cytokeratin, possibly after keratinocyte death.

Clinically, amyloidosis cutis dyschromica must be distinguished from xeroderma pigmentosum, dyschromatosis universalis hereditaria, dyschromatosis symmetrica hereditaria and dyskeratosis congenita.

Although the skin lesion develop extensively since childhood, systemic involvement is not evidence even after long-

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term follow-up. Amyloidosis cutis dyschromica with generalized morphea has been reported.

There are many special stains for amyloid include the triphenyl-methane dyes, methyl and cresyl violet for the demonstration of metachromasia, the periodic acid-Schiff method, the substantive cotton dyes Congo red and Sirius red with or without fluorescence or polarized light microscopy. Immunohistochemically, amyloid materials are stained by the polyclonal antikeratin antibody by using an avidin biotin peroxidase complex method.

Amyloidosis cutis dyschromica unfortunately responds relatively poorly to topical steroid, calcipotriol, phototherapy, and dermabrasion. There have been reports of response to etritinate or acitretin but not in all patients. Keratolytic agent may be useful.

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August 18, 2006