

Case Report

Amyloidosis cutis dyschromia: a case report

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Abstract Amyloidosis cutis dyschromia is a very rare variant of primary cutaneous amyloidosis clinically characterized by hyper- and hypopigmented/depigmented macules in generalized distribution. Diagnosis is based on histopathological demonstration of amyloid deposits in papillary dermis and its confirmation by Congo red staining. We hereby report a case of amyloidosis cutis dyschromia.

Key words

Amyloidosis cutis dyschromia, hyperpigmentation, hypopigmented macules, amyloid deposits.

Introduction

Amyloidosis cutis dyschromia is a very rare variant of primary cutaneous amyloidosis. It was first described by Morishima in 1970. It is characterized by generalized, mottled, hyperpigmented, hypopigmented and/or depigmented macules in generalized pattern with no/little pruritus, prepubertal onset and focal amyloid deposition in papillary dermis. Here we report a case of amyloidosis cutis dyschromia with positive family history.

Case report

A 30-yrs-old male presented with generalized mottled hyperpigmented and hypopigmented macules over trunk and both upper and lower extremities since 12 years of age. Initially the hypopigmented macular lesions appeared on his trunk and proximal part of upper and lower limbs, then gradually involved the distal

extremities with sparing of face, palm, soles, hands, feet, nails, teeth and mucosa. Patient did not complain of pruritus or photosensitivity. Erythema, telangiectasia or atrophy was absent. There was no history of extensive sunlight exposure, blisters, any inflammatory cutaneous conditions or systemic illness prior to the onset of lesions. Also there was no history of delay of developmental milestones.

He was born to nonconsanguineous parents. His younger sister, 25-year-old, had similar speckled spotty hyper- and hypopigmented macular lesions over trunk and extremities since 8 years of age (**Figure 1**).

On general physical examination, patient was normal and healthy. His height was 172 cm. Systemic examinations were not remarkable. On cutaneous examination, there were multiple hyperpigmented, hypopigmented and depigmented macules of size ranging from 1 mm to 15 mm involving trunk, buttocks, arms and forearms, thighs and legs sparing face, palms, soles, hands, feet, nails, teeth and mucosa (**Figure 2**).

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For histopathological examination, biopsy sample was taken from sun-protected site i.e. lower back, by punch biopsy, stained with hematoxylin-eosin stain and seen under light microscopy. It demonstrated small, globular, pink, amorphous amyloid deposits and melanophages in papillary dermis (**Figure 3**) These amorphous deposits were further stained with Congo red stain which was positive (**Figure 4**). In epidermis compact and orthokeratosis and hyperplastic rete ridges were seen. Occasionally, dyskeratotic cells were seen in the upper layers of epidermis. No changes were revealed in reticular dermis.

All laboratory parameters were normal on routine investigations.

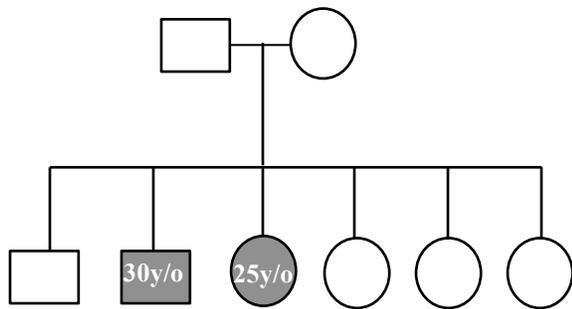


Figure 1 Patient's family tree.



Figure 2 Depigmented and hyperpigmented macules on back.

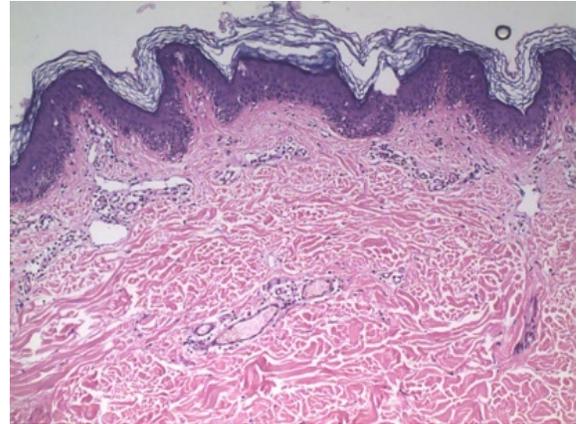


Figure 3 Hematoxylin and eosin staining showed hyperplastic rete ridges, compact, orthokeratosis, globular, pink, amorphous, amyloid deposits and occasional melanophages in papillary dermis.

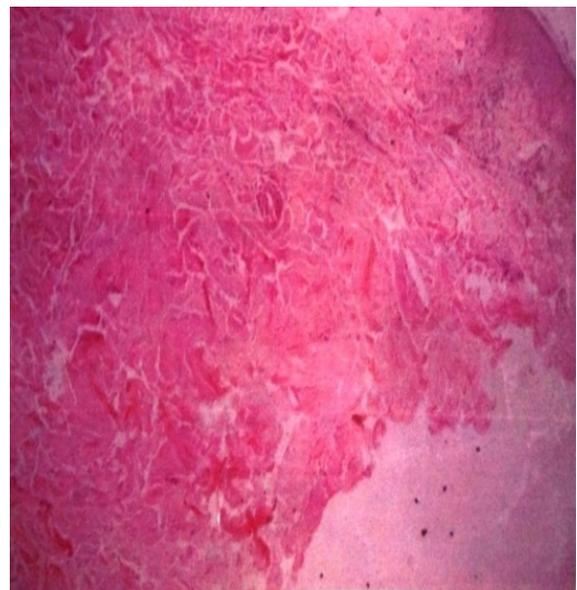


Figure 4 Eosinophilic deposits showing positive reaction with Congo red stain.

Based on clinical features and histopathological demonstrations, diagnosis of amyloidosis cutis dyschromia was made and patient was advised for oral acitretin and periodic follow-up.

Discussion

Primary cutaneous amyloidosis has been classified into three major types: lichenoid, macular and nodular (rare). Other atypical variants has also been described which are dyschromic amyloidosis or amyloidosis cutis

dyschromica, bullous, vitiliginous, familial poikiloderma like cutaneous amyloidosis and anosacral type. Amyloidosis cutis dyschromica is a very rare variant of primary cutaneous amyloidosis which was first described by Morishima in 1970. Till date, less than 30 cases of amyloidosis cutis dyschromica have been described in medical literature.

It is characterized by following features:

1. Mottled hyper- and hypopigmented macules
2. Onset before puberty
3. Usually non pruritic
4. Amyloid deposition in papillary dermis

Amyloidosis cutis dyschromica is assumed to be a familial disease with sunlight exposure as a major etiological factor. Its pathogenesis is unknown. In genetically predisposed individuals UVB and UVC rays leads to defective DNA repair and apoptosis of keratinocytes. These apoptotic keratinocytes gets phagocytosed by histiocytes or fibroblasts. The cytokeratins of lysed keratinocytes give rise to the amyloid material which gets deposited in the skin.

There are also few other cutaneous dyschromic diseases which must be ruled out for the clinical diagnosis of amyloidosis cutis dyschromica. They are poikiloderma like amyloidosis, xeroderma pigmentosum, dyschromic universalis hereditaria (DUH), acral reticulate pigmentation of Dohi, chronic radiodermatitis, dyskeratosis congenita, arsenical hyperkeratosis and drug-induced dyschromia due to mono-benzyl ether of hydroquinone, tetracycline, DPCP, thiazides and afloqualone. Poikiloderma like amyloidosis is differentiated by the presence of photosensitivity, blisters, palmoplantar keratoderma, lichenoid papules and short stature.

Amyloid deposits are absent in DUH, DSH and xeroderma pigmentosum.

Various treatment modalities have been described. Protection from sun exposure and use of sunscreen is advised to all the patients. Variable results have been seen with topical corticosteroids, keratolytics, dimethyl sulfoxide, oral vitamin C and E and CO₂ laser. Systemic retinoic acid derivatives has been very effective

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