*Journal of Pakistan Association of Dermatologists*. 2015;**25 (4)**:319-321.

**Case Report**

# Amyloidosis cutis dyschromica

**Ranju Choudhary, Madhu Sudhanan V, Mritunjay Kumar, Shyam Sundar Chaudhary**

Department of Skin, VD & Leprosy, Rajendra Institute of Medical Sciences, Bariatu, Ranchi

**Abstract** Amyloidosis cutis dyschromica (ACD) is a rare form of cutaneous amyloidosis, characterized by generalized, asymptomatic hyperpigmentation intermingled with several hypopigmented spots without papulation, atrophy and telangiectasia. Its onset usually begins before puberty. We hereby describe two female siblings of ACD belonging to tribal groups with no systemic involvement. This condition should be considered as a separate entity and need to be differentiated from other variants of primary cutaneous amyloidosis.

***Key words***
Amyloidosis cutis dyschromica, amyloid, pigmentary disorder, Congo red.

## Introduction

Amyloidosis cutis dyschromica (ACD) is a very rare form of primary cutaneous amyloidosis (PCA) with very few cases reported in literature. Cutaneous amyloidosis has been classified as primary cutaneous amyloidosis (PCA), secondary cutaneous amyloidosis and systemic cutaneous amyloidosis. PCA is deposition of amyloid in a previously apparent normal skin with no apparent systemic involvement.[1-4]

ACD is a rare distinct variant of primary cutaneous amyloidosis.

## Case Report

A 30-year-old female born to nonconsanguineous parents presented with progressive and asymptomatic mottled hyperpigmentation involving almost the whole body, present since 8 years of age. The lesion appeared initially on her trunk. The hyperpigmentation extended gradually over the years to involve her extremities. She also noticed spotty hypopigmentation along with the hyperpigmented macules. The skin lesions were more pronounced over the trunk (**Figure 1**), back of the neck, upper limbs. There was no history of photosensitivity or extensive sun exposure.

On physical examination she was well nourished and her stature was 158 cm. Her teeth, hair, nail and mucosa of the mouth, palms and soles and vulva were normal. There was no hearing difficulty. Systemic examination was unremarkable. The mental and developmental milestones of the patient were normal. There were no any other inflammatory skin diseases.

Her 35-year-old sister has remarkably similar cutaneous manifestations hyperpigmentation associated with spotted hypopigmentation on the whole body from the age of 10 years. Other family member had no systemic manifestation (**Figure 2**).

The results of skin biopsies taken from the trunk in both patients were similar. The pathological section with hematoxylin and eosin stain revealed superficial perivascular infiltrate of melanophages. The papillary dermis shows a

**Address for correspondence**
Dr. Ranju Choudhary
Department of Skin, Venereology & Leprosy
RIMS, PO Bariatu, Ranchi -834009
Jharkhand, India
Email: ranju.choudhary82@gmail.com

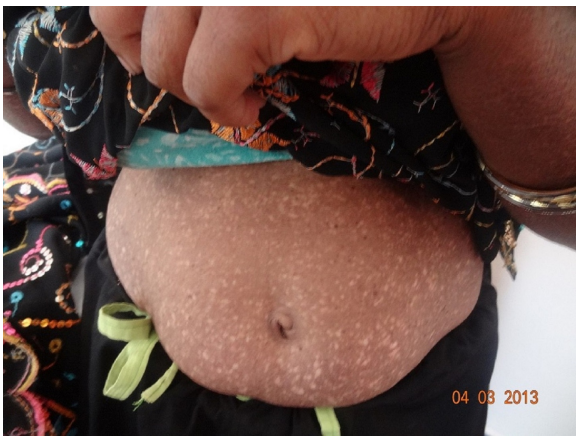


**Figure 1** The clinical features of the patient with hyperpigmented and hypopigmented macules on trunk.

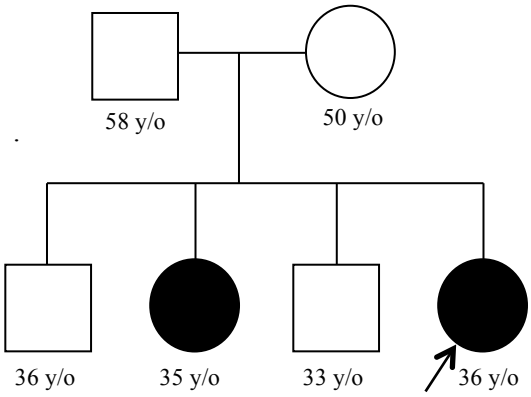


**Figure 2** Patient's family tree.

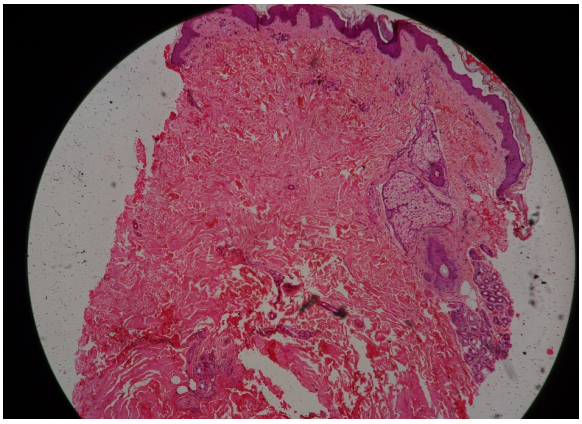

**Figure 3** The histopathological examinations of the patient with ACD. (A) H&E staining indicated hyperkeratotic epidermis and amorphous eosinophilic masses in the papillary dermis (original magnification × 40).

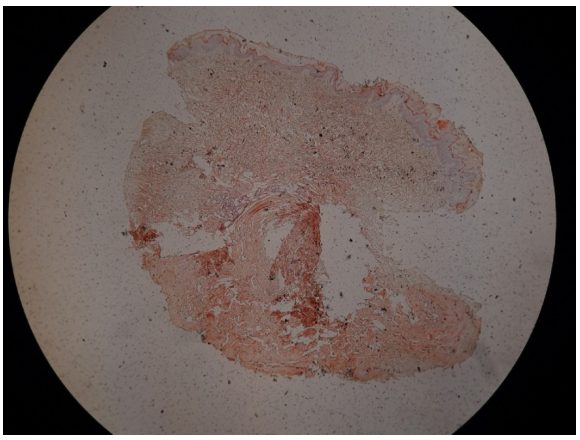

**Figure 4** Congo red staining was positive for eosinophilic masses in the papillary dermis (original magnification × 40).

few widened papillae. Some of these have small globular pink amorphous deposits of amyloid in association with melanophages (**Figure 3**). The Congo red staining was positive to eosinophilic masses (**Figure 4**), which indicated a deposit of amyloid substance.

Other results of routine laboratory examinations including routine blood test, biochemical profile, urine protein excretion and chest X-ray, abdominal ultrasound were normal

Based on the clinical and pathological findings, diagnosis of amyloid cutis dyschromica was made.

## Discussion

PCA includes the more common, papular variety (lichen amyloidosis), macular variety and nodular or tumefactive form.[2] ACD uncommon subtype of PCA was initially described by Morishima in 1970.

This disorder is characterized by the following features: (i) dotted or reticular hyperpigmentation with hypopigmented spots without papulation almost all over the body; (ii) no or little itchy sensation; (iii) onset before puberty; and (iv) small foci of amyloid closely

under the epidermis.[3,4] Our cases presented with reticular hyperpigmentation with hypopigmented spots, which were generalized distribution and onset before puberty. The amyloid deposits were limited to the subepidermal region, without other cutaneous and systemic involvement. All of these were ranged ACD.

Besides ACD, there are many other diseases with the characteristics of cutaneous dyschromia, includes dyschromatosis universalis hereditaria, xeroderma pigmentosum and poikiloderma-like amyloidosis. It is necessary to differentiate these diseases.[4,6-8] Amyloid deposits cannot be found in the skin of the patients with dyschromatosis universalis hereditaria and xeroderma pigmentosus. ACD and poikiloderma-like cutaneous amyloidosis have similar clinical features. Poikiloderma-like cutaneous amyloidosis is characterized by the presence of poikiloderma lesions, lichenoid papules and blisters especially located on the limbs and appears in adult life. It is often associated with photosensitivity, low height and a certain degree of palmoplantar keratoderma. A few clinical features are overlapping between ACD and poikiloderma-like amyloidosis. In our patients, there is mottled hypopigmentation and hyperpigmentation of skin but no telangiectasia and palmoplantar keratoderma. Also their statures were normal. So this is clearly a case of amyloidosis cutis dyschromica.

The etiology of PCA remains unknown, but it is believed to be multifactorial. Our patients are two female siblings. Owing to the familial aggregation, it suggests that genetic factors do play an important role in its pathogenesis. Vijaikumar *et al.*[5] proposed that genetic factors may lead to UVB sensitivity and DNA repair defects. This repeated damage to the keratinocytes results in the production of amyloid materials in the skin.[2]

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