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Original article

Clinical and histopathological study of primary cutaneous macular amyloidosis

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Abstract

Primary cutaneous amyloidosis often presents with pigmentary dystonias of the skin in the form of asymptomatic reticulate hyperpigmentation or pruritic lichenoid papular lesions. The aim of this study was to evaluate the incidence of primary cutaneous macular amyloidosis and also to find out the possible etiological agents, to correlate their clinical disease with histopathological positivity for amyloid deposition, and to find out the percentage of positive cases by special stains. A total of 24 patients attending dermatology out-patient clinic of Princess Esra Hospital, Hyderabad over a period of 1 year presenting with hyperpigmented skin lesions and clinically diagnosed as macular amyloidosis were taken up for this study.

Key words: Cutaneous amyloidosis, macular amyloidosis, Congo red stain

The generic term amyloidosis signifies the abnormal extracellular tissue deposition of one of the family of biochemically unrelated proteins that share certain characteristic staining properties including fibrillar ultrastructure and apple green birefringence of Congo red stained preparation viewed under polarized light[1]. Amyloid is a proteinaceous substance that is deposited in tissue in a wide variety of conditions. A number of stimuli can induce its formation and it can occur in association with many disorders. It can often be recognized in histological sections stained with hematoxylin-eosin as homogenous faintly eosinophilic aggregates that contain clefts as a result of shrinkage of amyloid during fixation and dehydration. Three staining methods are used for the demonstration of amyloid—Crystal violet, Congo red and Thioflavin T[2]. The most reliable method is staining paraffin embedded sections with Congo red and studying under polarized light. The amyloid shows green birefringence[2].

In systemic amyloidosis deposition of amyloid occurs in many organs whereas in localized amyloidosis it is restricted to single tissue or organ. In both the conditions there is considerable associated tissue dysfunction.

Primary cutaneous amyloidosis is a localized type of amyloidosis where amyloid is deposited in the skin and occurs as three variants—macular amyloidosis, lichen amyloidosis and nodular amyloidosis. Light microscopy findings indicate that in macular and lichen amyloidosis the degenerating epidermal cells are discharged into the dermis where they are converted to amyloid. These cells resemble colloid bodies and contain lysosomes and tonofilaments[3]. It is assumed that the degenerating tonofilaments are recognized as foreign and are digested by the cells' own lysosomes by conversion of pleated sheet configuration of the tonofilament to the β configuration of amyloid[2]. The epidermal derivation of amyloid in macular and lichen amyloi-

dosis is supported by histochemical and immunological findings, in contrast to the systemic amyloidosis. The amyloid in macular and lichen amyloidosis shows fluorescence for disulfide bonds as normally seen in stratum corneum and reacts to anti human keratin antibodies[2].

Etiological factors associated with primary cutaneous amyloidosis include racial susceptibility to lichen amyloidosis in people of the middle east, Asia, central and south America; and its striking familial presentation suggests the role of genetic factors.

Primary cutaneous amyloidosis is seen in relation to chronic friction such as use of nylon brush, nylon towel, etc. It is very likely that strong unbreakable nylon fibres or bristles abrade the stratum corneum damaging the keratinocytes. The degenerated keratinocytes maybe converted to amyloid at the level of dermoepidermal junction[4-7]. Primary localized cutaneous amyloidosis has also been found associated with long wave ultraviolet therapy used for treatment of psoriasis.

Materials and Methods

A total of 24 patients attending the dermatology clinic of Princess Esra Hospital, Deccan College of Medical Sciences, presenting with hyperpigmented skin lesions and clinically diagnosed as macular amyloidosis, were taken up for the present study.

A detailed history regarding the duration of disease, family history, and use of cosmetics, rough scrubbers, and massage oils was taken. All the patients underwent a thorough cutaneous and physical examination. No patient had evidence of systemic amyloidosis. They were subjected to routine investigations of blood, urine and stool and liver function and renal function tests, etc. A skin biopsy was performed and stained with hematoxylin-eosin and Congo red stains.

Results

The data obtained is tabulated according to various parameters like age, sex, possible etiological factors, symptoms, pattern of pigmentation, site of involvement, Congo red positivity and histopathological spectrum (Tables 1-5). Taking an overall perspective of the results obtained from the study following conclusions can be drawn.

1. Relatively young patients with cutaneous amyloidosis seek treatment, than older age groups.
2. Though friction plays an important role in the genesis of cutaneous amyloidosis (58.3%), a considerable percentage of patients (41.6%) did develop it in absence of friction, suggesting other factors.
3. Histopathological study of the biopsy specimens, revealed Congo red positivity of amyloid in 54.16% of cases and negativity in 45.33% of cases. This raises a doubt whether Congo red is sensitive enough to spot the minimal deposition of amyloid in the biopsy specimen. It was also revealed that popular lesions of cutaneous amyloidosis were associated with basal cell degeneration.
4. Cutaneous lesions found were predominantly diffuse pigmentation in 79.66% and rippled pigmentation in 58.33% of patients.

The present study revealed the common finding of deposition of eosinophilic hyaline material in papillary dermis in 62.5% and in dermal papilla in 25% of patients. Melanin incontinence and presence of melanophages in the dermis was seen in 16.66% of patients. Non-specific inflammation was present in another 16.66%. Acanthosis, hyperkeratosis and basal cell degeneration was seen in 4.16, 12.5 and 8.33 percent of patients respectively.

Table 1: Age wise distribution of cases

Age (years)	No. of patients	% of patients
15-24	11	45.83
25-34	7	29.16
35-44	3	12.5
45-55	2	8.33
55-64	1	4.16
Total	24	100

Table 2: Gender distribution of patients

Sex	No. of patients	% of patients
Males	4	16.6
Females	20	83.3
Total	24	100

Table 3: Etiological factors

	No. of patients	% of patients
Friction-Pumice stone	2	8.33
Friction-Nylon brush	12	50
No history of friction	10	41.70
Total	24	100

Table 4: Types of skin lesions

Pattern of pigmentation	No. of patients	% of patients
Discrete macules	11	45.83
Diffuse pigmentation	19	79.16
Rippled pigmentation	14	58.33
Lichenoid lesions	2	8.33

Table 5: Histopathological spectrum

Histological features	No. of patients	% of patients
Acanthosis	1	4.16
Hyperkeratosis	3	12.5
Basal cell degeneration	2	8.33
Deposition of eosinophilic hyaline papillary tips	6	25
Deposition of eosinophilic hyaline papillary dermis	15	62.5
Melanin incontinence	4	16.66
Non-specific inflammation	4	16.66

Discussion

The present study included 24 patients diagnosed clinically as having cutaneous amyloidosis. An attempt was made to find out the possible etiological factors and to study the clinical and histopathological spectrum of the disease.

As indicated in table 1, the incidence of macular amyloidosis is 87.4% in the age group of 15-44 years.The fact that cutaneous amyloidosis is more common in females is well established which can be seen in table 2.

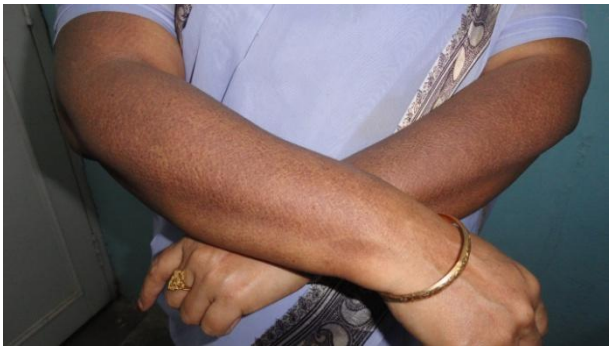

Fig 1a. Lesion on extensor aspect of forearms

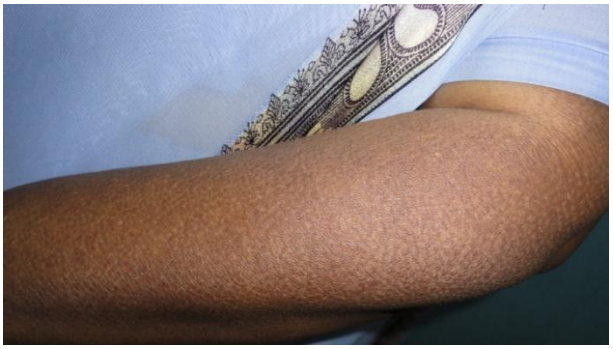

Fig 1b. Lesion on extensor aspect of forearm

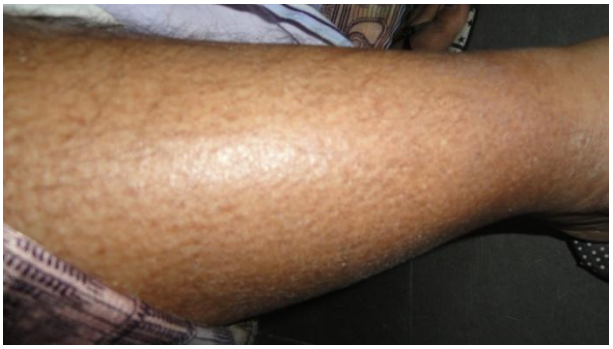

Fig 2. Lesion on shin

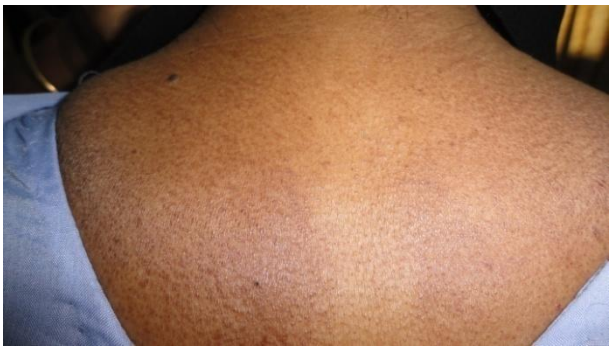

Fig 3. Lesion on upper back

Keeping in mind the objectives of finding the possible etiological factors such as the use of rough surface object for scrubbing the body while bathing, it was found that 56.33% of patients gave a positive history of using nylon scrubber (Table 3). The nylon scrubbers and towels on long term use

cause macular amyloidosis over the skin and bony prominences of upper part of torso and anterior aspect of legs[7].

Phototherapy using psoralen and long wave ultraviolet light was also reported to be one of the causes of macular amyloidosis. A study on 52 patients receiving PUVA therapy by Greene et al found that after about 1-2 years, 6 of them developed amyloidosis and demonstrated superficial deposit of amyloid in skin[8].

It can be hypothesized that chronic sun exposure leads to cutaneous amyloidosis. This is supported by the fact that in our study all the patients had lesions on extensor aspects of forearm (Fig 1a and 1b), shin (Fig 2) and 54.1% on upper back (Fig 3) which are mostly exposed to sunlight in common Indian dress code.

The clinical spectrum of pigmentation as shown in table 4 was predominantly of diffuse (79.16%) and rippled (58.33%) pattern. Discrete macules were seen in 45.8% and lichenoid lesions in 8.3% of patients.

The paucity of histopathological changes in epidermis has been uniformly mentioned in the literature. In the present study 25% of the cases had eosinophilic hyaline material in dermal papillae and 62.5% in papillary dermis (Table 5).

It is assumed that damage to epidermis is an essential feature which occurs prior to amyloid deposition. Amyloid if present in large amount in the skin sections can be recognized in hematoxylin-eosin stain but the most reliable method for its demonstration is staining with alkaline Congo red, where amyloid gives green birefringence[9].

Conclusion

We infer that, cutaneous amyloidosis is seen more commonly among younger patients particularly females, with common sites of involvement being the extensor aspects of forearms, upper back and shins.

Most of the patients reported hyperpigmentation of diffuse or rippled type. Pruritis was not major accompanying symptom. In the Indian context, friction forms a major etiological factor but other factors like chronic sun exposure and other climatic conditions are contributory.

The major histological change was deposition of eosinophilic hyaline material in dermal papilla and papillary dermis. Other features seen less frequently were hyperkeratosis, acanthosis, basal cell degeneration and presence of melanophages.

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Conflict of interest: None

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