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CLINICO-EPIDEMIOLOGICAL PROFILE FOR PRIMARY CUTANEOUS AMYLOIDOSIS IN A TERTIARY CARE CENTRE OF NORTH INDIA.

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ABSTRACT **Background:** Primary cutaneous Amyloidosis (PCA) is a skin disorder associated with the amyloid accumulation extracellular space. This study aimed to identification of clinico-epidemiological parameters of PCA. **Methods:** A cross-sectional study was done on thirty-six consecutive patients with a clinical diagnosis of PCA made by two independent dermatologists. Clinico-epidemiological, parametres recorded for all patients. Archived images were reviewed by two independent dermatologists. **Results:** Out of 36 clinically diagnosed PCA patients have a predominance in middle-aged females (mean age 42.75 ± 10.55 years; male-to-female ratio 1:2.5) with Fitzpatrick skin types IV-V, commonly affecting the upper back (88.89%) in a rippled pattern (91.67%). The mean age at onset was 37.90 ± 10.55 years, with most patients (58.33%) presenting within 2 years (mean duration 3.91 ± 2.46 years) and reporting pruritus (94.44%). Family history was noted in only 19.44%. **Conclusions:** Among 36 clinically diagnosed PCA patients, middle-aged females predominated (mean age 42.75 ± 10.55 years; M:F ratio 1:2.5) with Fitzpatrick IV-V skin types. Upper back involvement (88.89%) showed rippled patterns (91.67%), with onset at mean of 37.90 ± 10.55 years, short duration (mean 3.91 ± 2.46 years), pruritus (94.44%), and family history in 19.44%.

KEYWORDS : Primary Cutaneous Amyloidosis, Clinical profile of PCA, Epidemiology of PCA

INTRODUCTION

Amyloidosis is a skin disorder characterised by the extracellular accumulation of amyloid which is a proteinaceous substance that is resistant to proteolytic processing, often leading to significant tissue dysfunction. There are two types of cutaneous amyloidosis, primary localised cutaneous amyloidosis (PCA), which does not involve internal organ deposits and secondary cutaneous amyloidosis, which affects multiple organs and tissues.

Talking about PCA, it is a common condition in the Asian population with an overall prevalence ranging from 0.03% to 1.13%in the Indian population.[1–4] The condition is most prevalent in individuals aged 21 to 50 years[5], particularly females with a female-to-male ratio varying from 1.16:1[6] to 3.16:1[7] as per different studies. Although the etiology of PCA is unknown, some minor associations have been found with friction, scrubbing, and exposure to sunlight.

ClinicalClassification:

Amyloidosis is categorised into two types: systemic and cutaneous. In cutaneous form, amyloid accumulation is typically found around the affected area, and in systemic type, it accumulates in the blood and is deposited in various organs.

Clinical Classification Of Amyloidosis [8]

I. Systemic Amyloidosis

1. Primary systemic amyloidosis
2. Secondary systemic amyloidosis
3. Heredo-familial amyloidosis

II. Organ-limited Amyloidosis

1. Cutaneous amyloidosis
2. Endocrine amyloidosis
3. Cerebral amyloidosis

Clinical classification Cutaneous Amyloidosis[8]

A. Primary Cutaneous Amyloidosis –

1. Macular amyloidosis (MA)
2. Lichen amyloidosis (LA)
3. Nodular amyloidosis (NA)

B. Secondary Cutaneous Amyloidosis

The disease has various clinical variants, including macular amyloidosis (MA), lichen amyloidosis (LA), nodular amyloidosis (NA), biphasic amyloidosis (BA), amyloidosis cutis dyschromica (ACD), and anosacral amyloidosis. MA is the most prevalent subtype, presenting as asymptomatic or pruritic brownish macules with a rippling or confluent pattern involving upper back (interscapular areas) and extremities (shins and forearms) while multiple pruritic, firm, hyperpigmented, hyperkeratotic papules on the shins in a rippling pattern are suggestive of LA. The coexistence of macular and papular lesions in a patient is called BA.[9] [10]

INVESTIGATIONS:

Investigations performed for the diagnosis include the following:

1. Staining
a. Haematoxylin and Eosin stain (H & E stain)
b. Special stains- Congo Red

2. Immunohistochemistry (IHC)
3. Electron-microscopy (EM) and Reflectance confocal microscopy (RCM)
4. X-ray crystallography and infrared spectroscopy
5. Direct Immunofluorescence (DIF)

Biopsy continues to be the preferred method for confirming the diagnosis of PCA with the presence of amyloid deposition in the papillary dermis being the characteristic histopathological finding.

This study focused on the identification of clinico-epidemiological parameters of PCA.

MATERIALS AND METHODS

Study Design:

This Prospective cross-sectional study was conducted in the outpatient department of Dermatology at our institute from December-2022 to December-2023. Thirty-six consecutive patients with a clinical diagnosis of PCA (made by two independent dermatologists, both sexes (males and females), aged >18 years, who have not taken any form of treatment in last 4 weeks for PCA and were willing to give written consent) were included in the study. Patients having rippled or confluent hyperpigmentation present over the upper back, arms, and legs were clinically diagnosed as PCA. Patients with concomitant systemic amyloidosis, pregnant or lactating women and immunocompromised were excluded from the study.

Study Sample Size:

We have included all patients came to our department within the study time period for the study database that fulfil the inclusion and exclusion criteria.

Data Collection And Analysis:

After a detailed history and clinical examination of the patients, clinical photographs were taken using a 108-megapixel (MP) mobile camera and archived.

Statistical Analysis:

Continuous data was expressed in mean+/-SD. Categorical parameters were expressed as frequency and percentage. The final analysis was done using Statistical Package for Social Sciences (SPSS) software,

version 25.0, IBM manufacturer, Chicago, USA.

RESULTS:

Total 36 patients are recruited from this time period. The age of the patients ranged from 21 - 76 years (mean age 42.75 ± 10.55 years). Out of 36 patients, 41.67% (n=15) belonged to the age group 21-40 years, and 44.44% (n=16) belonged to the age group 41-60 years. There were only 13.89% (n = 5) patients in the age group 61-80 years. In the present study, the male-to-female ratio was "1:2.5" with females accounting for 72.22% (n = 26). In the majority of patients, 58.33% (n =21) age at onset of disease was between 31-50 years of age followed by 25% patients (n = 9) in 11-30 years of age. The mean age at onset of disease was 37.90+ 10.55 years (ranging from 16 to 71 years). The lowest age at onset was 16 years and the highest age of onset was 71 years. The mean duration of diseases was 3.91 ± 2.459 (range = 1-10) years. Most of the patients 38.89% (n-14) presented within 2 years of the onset of diseases. Out of 36 patients, 94.44 % (n=34) patients reported itching or scratching. The family history of similar lesions was present in 19.44% (n = 7) patients. (Table-1)

The most commonly affected site was the upper back in 88.89% patients (n=32), followed by extensor surfaces of the arm in 58.33% patients (n = 21), extensor surfaces of the lower limb (pretibial) in 52.78 % (n=19) patients, and both, the forearm and the scapula were present in 38.89 % (n=14) patients each. The chest was involved in 13.89 % (n=5) patients, and the lower back in 11.11% (n=4) patients. Less commonly affected sites included the abdomen, which had lesions in 5.56%(n=2) patients and followed by both neck and thighs in 2.78% (n=1) patients. Simultaneous involvement of both upper back and extensor surface of the arm was present in 33.33% (n=12) patients. (Figure-1) Around 72.22% (n=26) of the patients had Fitzpatrick skin type 4 while 30.56% (n=11) patients had skin type 5. Most of the patients 75% (n=27) had macular presentation followed by papulo-macule in 16.67% (n=6) and the least commonly papules in 13.89% (n=5). Most of the patients had clinical presentation as rippled pattern in 91.67 % (n=33) and confluent pattern in 11.11 % (n=4) patients. Based on clinical presentation, 61.11% (n=22) patients had macular amyloidosis, 22.22% (n=8) patients had lichen amyloidosis, and 16.67% (n=6) patients had biphasic amyloidosis. (Table-1)

DISCUSSION

In our study among the 36 clinically diagnosed patients, the age of majority of patients was between 41-60 years (44.44%), followed by 21-40 years (41.67%). This is in concordance with other studies such as Biswas et al[3] which reported 46% presentation between 20 to 39 years. The mean age of patients was 42.75 ± 10.55 years (range = 21 to 76 years) it has accordance with a study by Mehrotra et al[11] (36 + 11.7 years). In various literatures, female predominance was reported[3,12,13] also in our study, the male-to-female ratio was 1:2.5 with females accounting 72.22% (n = 26). In the majority of patients, the age of disease onset was between 31-50 years (58.33%) of age followed by 11-30 years (25%). The mean age at onset was 37.90+ 10.55 years, ranging between 18 to 71 years of age, that is in concordance with other studies by Biswas et al[3] (38.35+ 13.7 years), Bandhlish et al[1] (34.6+10.5 years). The mean duration of diseases was 3.6 ± 2.2 years (ranged=1-10) years. Out of 36 patients, 94.44 % patients reported itching or scratching. According to the literature, most of the studies reported it between 34 to 68% patients[1–3,12–15] only one study done by Salim et al[16] reported it around 90 % which is comparable to our finding.[16] There is less data on family history of PCA, only some studies reported it between 10 to 20 % of patients in their studies.[2,3,14,15] In our study, the family history was present in 19.44 % patients.

In literature most commonly involved sites were upper back,[1,2,15,17] extensor surfaces of arm,[14,18] and extensor surfaces of lower limb (pretibial)[3,12,13,16]. In our study also, the most commonly affected site was the upper back 88.89%, followed by extensor surfaces of arm 58.33%, extensor surfaces of lower limb (pretibial) 52.78 %, forearm and the scapula each in 38.89% patients. The chest was involved in 13.89% patients and the lower back in 11.11% patients. Less commonly affected sites included the abdomen 5.56%. Simultaneous involvement of both upper back and extensor surface of arm was present in 33.33% (n=12) patients. In our study, around 72.22% (n=26) of the patients had Fitzpatrick skin type 4 while 30.56% (n=11) patients had skin type 5. Most of the patients in our study 75% (n=27) had macules followed by papulo-macule 16.67% (n=6) and least commonly papules in 13.89 % (n=5). In most of the available literature data, rippled pattern is most common clinical presentation reported.[1–3,14,17–20] In our study also, most of the patients had clinical presentation as rippled pattern (91.67%) with confluent pattern present in 11.11% patients.

CONCLUSION

This study delineates the clinical and epidemiological profile of 36 patients with rippled hyperpigmentation, revealing a predominance in middle-aged females (mean age 42.75 ± 10.55 years; male-to-female ratio 1:2.5) with Fitzpatrick skin types IV-V, commonly affecting the upper back (88.89%) in a rippled pattern (91.67%). The mean age at onset was 37.90 ± 10.55 years, with most patients (58.33%) presenting within 2 years (mean duration 3.91 ± 2.46 years) and reporting pruritus (94.44%). Family history was noted in only 19.44%.

Table-1: **Clinico-Epidemiological Characteristics Of PCA**

S. No.	Characteristics	Mean SD % (n), (n = 36)	
1.	Age of onset of PCA (years)	37.90+ 10.55	
2.	Age at enrolment in the study (years)	42.75 ± 10.55	
3.	Duration of disease (years)	3.6 ± 2.2	
4.	Male:Female	1:2.37 (n=8/19)	
5.	Itching/scratching	94.44 % (n=34)	
6.	Family history	19.44% (n = 7)	
7.	Site of involvement	Upper back Extensor surfaces of the arm Extensor surfaces of lower limb (pretibial) Extensor of forearm Scapula Chest Lower back Abdomen Neck Thigh	88.89% (n=32) 58.33% (n = 21), 52.78 % (n=19) 38.89 % (n=14) 38.89 % (n=14) 13.89 % (n=5) 11.11% (n=4) 5.56%(n=2) 2.78% (n=1) 2.78% (n=1)
8.	Clinical patterns	Rippled	91.67 % (n=33)
		Confluent	11.11 % (n=4)
9.	Type of primary cutaneous amyloidosis	Macular amyloidosis	61.11% (n=22)
		Lichen amyloidosis	22.22% (n=8)
		Biphasic amyloidosis	16.67% (n=6)

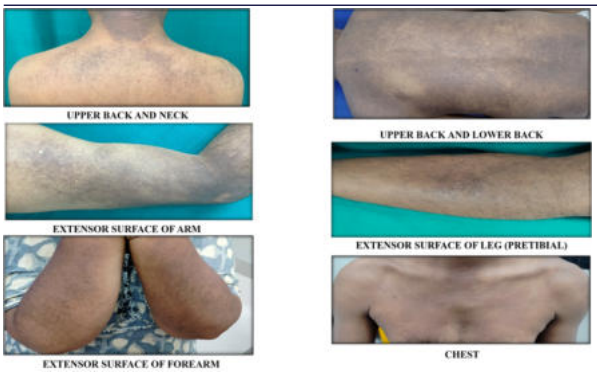

Figure-1: Clinical Pictures of PCA at Various Sites

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