Dermoscopy of Macular Amyloidosis

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Case 1: A 47-year-old Indian lady with controlled hypothyroidism and Fitzpatrick skin phototype (SPT) IV presented with 3-years-old macular hyperpigmentation in a rippled pattern over her upper back [Figure 1a], and signs associated with mild pruritus.

Case 2: A 39-year-old Indian man with SPT IV presented with more than 10-years-old macular hyperpigmentation in a rippled pattern over the outer aspect of his arms and legs [Figure 1b]. The pruritus had a fluctuating course, with worsened itching in the past 3 months being his major presenting complaint.

Past history of chronic rubbing was negative in both cases. Family history of similar pigmentation was present in both parents of the male patient. Excepting moisturizers and occasional oral antihistamines, neither patient had used any other treatment in the past 6 months.

On polarized videodermoscopy [Escope videodermoscope, 20×] from randomly selected areas of the involved skin (avoiding scratched/modified areas) in both cases [Figure 2a and b from the back of case 1 and Figure 2c and d from the leg of case 2], multiple features and pigmentary patterns were observed in all images, with relative predominance of certain features in some [Table 1]. Histopathology with hematoxylin and eosin, and special staining with Congo red confirmed the diagnosis of macular amyloidosis in both cases with the presence of small globular pink amorphous deposits of amyloid in the papillary dermis, lamellated irregular orthohyperkeratosis (more in Case 1), mild parakeratosis, and dermal melanophages (more in Case 2). The spectrum of dermoscopic features and their *postulated* histogenesis are detailed in Table 1.

Macular amyloidosis (MA) a common form of primary cutaneous amyloidosis (PCA) typified by itchy brownish macules in a rippled pattern is very common in Asians, especially Taiwanese and Indians.[1] Although MA is not classified as a "pigmented disorder" *per se*, hyperpigmentation predominates clinical presentation. The diagnosis is usually clinical and straightforward. However, differentiation from conditions like lichen planus pigmentosus (LPP), lichen simplex chronicus, post-inflammatory hyperpigmentation, poikiloderma of Civatte, and poikiloderma-like cutaneous amyloidosis may be occasionally needed. In the only thoroughly conducted published study on dermoscopy of MA Chuang *et al.* in 18 Taiwanese individuals, central hub(s) of brown, white or both colors constituted the primary feature, surrounded with brownish pigmentation that was classified into three main patterns – fine streaks ("spoke-wheel" pattern), leaf-like extensions, and thick noncircular pedal projections.[2] Although the SPTs of the included Taiwanese patients were not specified in this study, other published sources suggest that Taiwanese people typically have SPT type III–V.[3] Further, the published indexed literature on dermoscopy of MA in patients with skin of color (SOC) is scarce, excepting for the singular case report by Kaliyadan *et al*. on dermoscopic features of PCA (biphasic) described in a 57-year-old Indian woman with SPT V.[4] While authors observed areas corresponding to the pattern described by Chuang *et al*. in the form of white hubs surrounded by different patterns of hyperpigmentation, the specific morphology and amount of pigmentation were different. The pigmentation was darker and formed thicker structures. In particular, radial streaks and venation were not seen.[4] It is noteworthy that

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Table 1: The diverse dermoscopic features observed in macular amyloidosis in two Indian patients, with some histopathological correlates

Dermoscopic Feature	Images showing it as a conspicuous feature	Hypothesized histopathological correlate
Multiple uniform small central hubs with ill-defined and broken brown streaks radiating from the center giving a "hub-and-spoke" pattern (white and yellow circles). Radial streaks were not regularly seen. Multiple hubs-and-spokes in a single dermoscopic field were of common occurrence. The color of the central hub was primarily off-white (white circles), followed by light brown (yellow circles), and in certain images it showed a "brown-in-white" appearance (yellow circles)	Figure 2a-d	The color of the central hub depends on the extent of hyperkeratosis appearing more white in cases with marked hyperkeratosis [Figure 2a], and brownish if hyperkeratosis is milder with only loose, basket-weave orthokeratosis [Figure 2d]
Lode-like* pigmented dark brown to almost black structures (blue arrows) in proximity with a central hub	Figure 2b-d [Most prominent in 2b]	Basal hyperpigmentation, pigment incontinence and melanin granules within papillary dermal amyloid deposits
Lode-like* pigmented dark brown to almost black structures (blue arrows) forming bizarre patterns with no apparent topographical proximity with a hub	Figure 2b and c	Basal hyperpigmentation, pigment incontinence and melanin granules within papillary dermal amyloid deposits#
Scattered leaf venation-like pigmented structures in various configurations (blue circles)** - arcuate, palmate, reticulate, rotate, parallel etc.	Figure 2a-d (most appreciable in 2d)	
White-to-brown isolated hubs (red circles) independent of pigmented structures emanating from them or surrounding them	Figure 2b-d	Probably similar to the hubs described in S no. 1
Perifollicular depigmented halos (yellow arrows)	Figure 2a-c	Could be similar to the "holes" seen as a part of pseudonetwork; where pigment network is absent due to adnexal openings such as hair follicles in this case
White-to-off white colored intersecting lines and/or white-to-light brown colored structureless areas	Figure 2a-d	Structureless areas may occur due to irregular hyperkeratosis thereby obscuring underlying structures in areas with thicker stratum corneum
Focal fine white non-adherent scales (black arrows)	Figure 2a-c	Most likely suggestive of focal parakeratosis

*'lode' is a deposit of metalliferous ore that fills or is embedded in a fissure in a rock formation. - Figure 2b [Inset] demonstrates the morphology of lode-like deposits. ** Figure 2d [Inset] schematically demonstrates the diverse morphology of leaf-venation patterns. #The exact explanation of the varied configurations and patterns of pigmented structures and their presence in proximity of the hubs as well as discreet scattering needs to be studied with a clinico-dermoscopic-pathological study including special stains

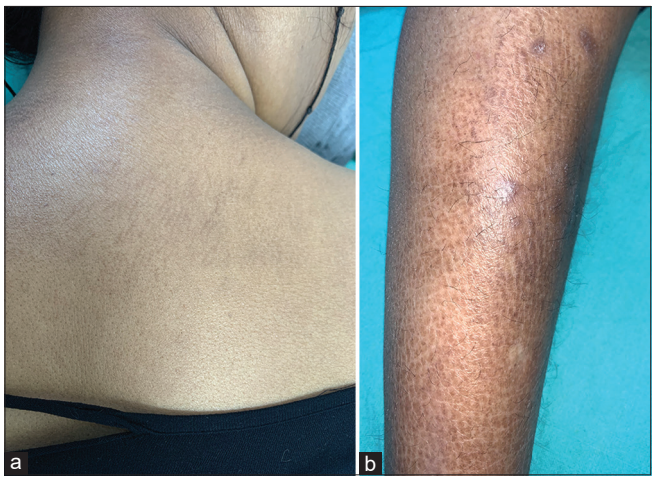

Figure 1: Clinical images of two patients with macular amyloidosis: (a) Case 1 - Rippled macular pigmentation over the upper back of an Indian lady; and (b) Case 2 - Similar macular pigmentation over the leg of an Indian man. Excoriation marks and superficial scales are also visible

streaks were similarly not well-visualized in our cases too. Even when seen, they were irregular and often broken [Figure 2a]. Moreover, the pigmentation was also darker and formed thicker lode-like structures [Figure 2b]; although leaf venation structures were seen in case 2. Although based on our two cases and the case by Kaliyadan *et al*., it seems that dermoscopy patterns for primary cutaneous amyloidosis *may be* different in patients with darker skin types, the lack of information on SPT of Chuang *et al*.'s 18 cases, and scarcity of dependable cases reported from India render this presumption premature. The dermoscopic changes (especially pigment color), observed from 18 Taiwanese vs 3 Indian patients may at best be ascribed to the reported phenomenon of more extensive and darker pigment structures seen in dermoscopy of hyperpigmentary disorders in darker skin types.[5]

The histogenesis of the dermoscopic findings of MA remains unclear. Brown pigmentation on dermoscopy has been attributed to basal hyperpigmentation, pigment incontinence, and melanin granules within papillary dermal amyloid deposits.[2] The color of the

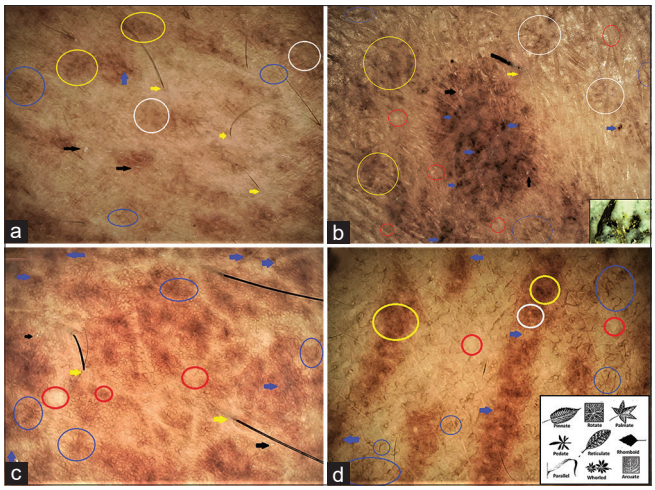

Figure 2: Dermoscopic images from the patients. Images A and B are from the back of case 1 and C and D from the leg of case 2 (but the two images of each patient have been taken from different foci within the expanse of macular amyloidosis): (a) Multiple foci of small central hubs with irregular and broken streaks radiating from the center giving a "hub-and-spoke" pattern were seen. The color of the central hub was either off-white (white circled) or light brown or mottled whitish-brown (yellow circled). Perifollicular area showed reduced pigmentation with almost a light-colored halo (yellow arrows). Additionally, pigmented structures also visible forming dark brown-colored venation of various configurations (blue circles) simulating the arrangement of veins in different leaves, and as scattered lode-like deposits, both in proximity with a hub (blue arrows) and forming bizarre patterns with no apparent topographical proximity with a hub. Focal fine scales also visible (black arrows); (b) The dark and thicker lode-like deposits in proximity with a hub (blue arrows) as well as forming bizarre patterns are better visualized in this image. The figure inset demonstrates the morphology of lode-like deposits. Additionally, white-to-brown isolated hubs independent of pigmented structures emanating from them or surrounding them (red circles) are also appreciable. The pigmented structures irrespective of their orientation and pattern are interspersed with white to off-white colored intersecting lines and white-to-light brown colored structureless areas; (c) Prominent features in this image include multiple scattered pigmented venation-like structures (blue circles), and a background network of light brown colored structureless areas with intersecting lighter colored lines; and (d) This image shows most of the features, with majority of the pigmented structures clustered along broad, almost parallel sulci, and others visible as dark brown venation-like structures, and bizarre-shaped arciform lines arranged in a racemiform pattern and more conspicuous in the intervening lighter colored areas due to color contrast. The figure inset demonstrates the diverse morphology of leaf venation patterns that simulate the venation patterns observed in dermoscopy of macular amyloidosis [Escope videodermoscope, polarized, 20×]

central hub was suggested to depend on the extent of hyperkeratosis by Chuang *et al*.[2] appearing more white in cases with marked hyperkeratosis (as in Case 1), and brownish if hyperkeratosis was milder (as in case 2). We observed additional findings and have suggested plausible postulates (without specific correlative confirmation) [Table 1], but at this point it is almost impossible to suggest a precise histopathological correlation for the observed features. We expect to publish a larger clinico-dermoscopic-pathological study with statistical quantification of dermoscopic features and probable histological correlation very soon.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Krishna A, Nath B, Dhir GG, Kumari R, Budhiraja V, Singh K. Study on epidemiology of cutaneous amyloidosis in Northern India and effectiveness of dimethylsulphoxide in cutaneous amyloidosis. Indian Dermatol Online J 2012;3:182-6.
2. Chuang YY, Lee DD, Lin CS, Chang YJ, Tanaka M, Chang YT, *et al*. Characteristic dermoscopic features of primary cutaneous amyloidosis: A study of 35 cases. Br J Dermatol 2012;167:548-54.
3. Wang CC, Tang CH, Wang CY, Huang SY, Sue YM. Risk of skin cancer in patients on chronic haemodialysis: A nationwide, population-based study in Taiwan. Br J Dermatol 2016;175:1175-82.
4. Kaliyadan F, Alkhateeb A, Kuruvilla J, Swaroop K, Alabdulsalam AA. Dermoscopy of primary cutaneous amyloidosis in skin of color. Dermatol Pract Concept 2019;9:232-4.
5. Sonthalia S, Gupta A, Jha AK, Sarkar R, Ankad BS. Disorders of pigmentation. In: Lallas A, Errichetti E, Ioannides D, editors. Dermoscopy in General Dermatology. Boca Raton, FL: CRC Press; 2018. p. 282-94.