Dermatology Practical & Conceptual

Pigmented Macular Variant of Chronic Cutaneous Lupus Erythematosus: Clinico-dermoscopic Features of Three Patients With Skin of Color

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To the Editor,

Chronic cutaneous lupus erythematosus (CCLE) is characterized by the presence of well-defined erythematous scaly plaques with associated atrophy, scarring, telangiectasia, and pigmentary alteration. The pigmented macular variant is a less recognized morphological variant [1].

All 3 cases in our series were female with Fitzpatrick skin phototype V. They presented with multiple, ill-defined, light-to-dark brown macules of variable size, predominantly on the face and head. Associated scarring was noticed only in Case #1. The dermoscopic feature was dominated by follicular plugging and pigment structures like brown dots, globules, homogenous areas, brown to blue-gray peppering, and brown pigment network. The brown to blue-gray peppering showed a peri-appendageal accentuation and resulted in target-like structures (central white clod/eccrine opening surrounded by brown structureless area) and perifollicular gray ring. Case-1 showed also concentric structures as dark brown clod within a light brown clod (Figure 1 and 2). The clinico-dermoscopic details of the 3 cases are given in Table 1. Two 4 mm punch biopsies were done: one for histopathological examination and the other for direct immunofluorescence from hyperpigmented macules. The histopathology of all the cases was diagnostic of CCLE. On direct immunofluorescence, linear deposition of immunoreactant IgG and, or IgM were noted along the basement membrane zone in all three cases.

George et al in 1992 reported a hyperpigmented variant of discoid lupus erythematosus and advocated that asymptomatic chronic hyperpigmented macules should alert the clinicians to suspect the diagnosis [1]. Boyd delineated

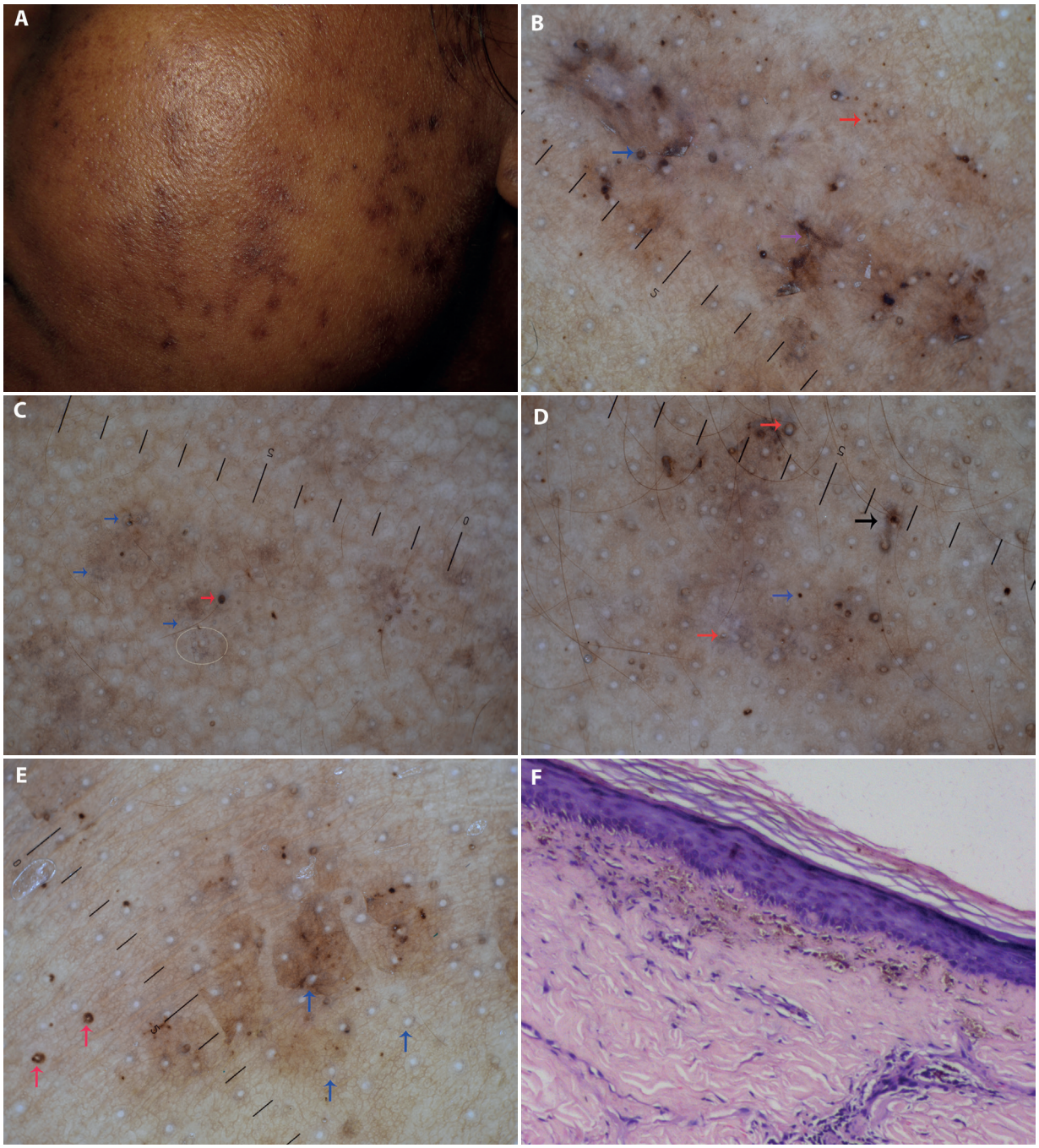

Figure 1. (A) Multiple ill-defined brown macules. (B) Dermoscopy under nonpolarized mode shows follicular plugging (blue arrow), brown dots, globules, and fine peppering (purple arrow), and concentric structures (red arrow), (C) Perifollicular gray ring (blue arrows), follicular plugging (red arrow), and perifollicular accentuated peppering (circle). (D) Brown globule (blue arrow), follicular plugging (red arrows), and perifollicular accentuated peppering (black arrow). (E) Peri-eccrine accentuation of peppering (blue arrows), and target-like structures (white clod surrounded by brown ring (red arrows). (F) Histology shows epidermal atrophy, melanin in stratum corneum, basal vacuolar degeneration, and pigment incontinence (H & E, x100).

a distinct profile of patients with solitary hyperpigmented macules over the face and neck without any scaling, atrophy, scarring, or telangiectasia [2]. These lesions had a late-onset, short disease duration, lower incidence of photosensitivity, and overall better prognosis. They could clinically be mistaken for seborrheic keratosis, solar lentigo or lentigo maligna, but the histopathology was consistent with CCLE. The three cases in our series had multiple asymptomatic light-to-dark brown macules without any previous history of inflammation and were distributed primarily on the face and around the ear.

The dermoscopic features, follicular plugging, brown to blue-gray dots, globules, or peppering in our series reflect the basic pathology of CCLE. The former corresponds to the dilated follicular infundibulum keratotic plugging, and the latter to the melanin in stratum corneum and to the dermal pigment incontinence. Besides, we observed perifollicular gray ring and peri-eccrine target-like structures, which

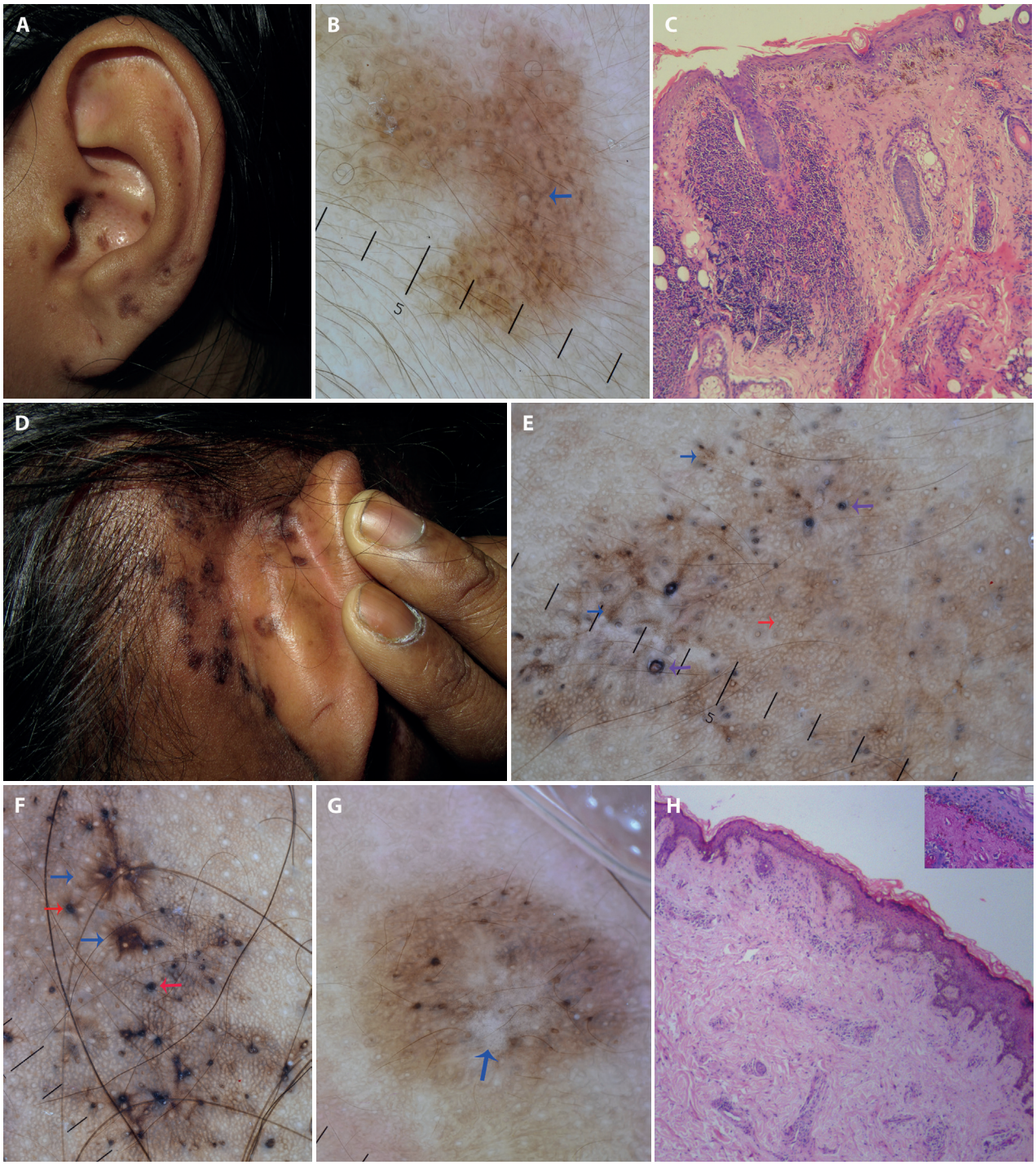

Figure 2. (A) Multiple ill-defined light brown macules on the ear. (B) Dermoscopy under nonpolarized mode shows blue-gray peppering (arrow) over a light brown background. (C) Histology shows epidermal atrophy, follicular plugging, interface dermatitis, and perifollicular lymphocytes and macrophages infiltration (H & E, x100). (D) Multiple ill-defined dark-brown macules over the retro auricular area. (E) Dermoscopy under nonpolarized mode shows follicular plugging (purple arrows), pigment network (red arrow), and peppering (blue arrows). (F) Brown pigment network, follicular plugging (red arrows), and periappendageal accentuated peppering (blue arrows). (G) Focal white homogenous area with overlying peppering (arrow). (H) Histology shows atrophic epidermis along with interface dermatitis (H & E, x100). Inset shows Periodic acid–Schiff positive thick basement membrane (PAS, x400).

possibly reflected the predominance of peri-appendageal interface dermatitis and the resulting pigment incontinence [3,4]. The pigmented macular variant of CCLE will have more prominent pigment incontinence in skin of color. This feature may not be striking in Fitzpatrick skin phototype I-II.

Diagnosing the pigmented macular variant of CCLE in skin of color, as in our patients, is challenging. It can mimic various acquired hyperpigmentation disorders such as lichen planus pigmentosus (LPP), pigment contact dermatitis (PCD), and melasma. Besides, it can also be confused

Table 1. Clinical and dermoscopic details of the pigmented macular variant of chronic cutaneous lupus erythematosus.

Cases	Gender/Age in years	Location	Duration in months	Associated features	Dermoscopic features
Case-1	Female/35	Face, upper trunk	Two	Nil	Brown dots, globules, and peppering Blue-gray peppering Follicular plugging Peri-appendageal accentuation of peppering Perifollicular gray ring Target-like structures Pseudo-pigment (brown) network Concentric structures
Case-2	Female/30	Bilateral ear and infra-auricular area	Three	SLE Non-scarring alopecia ISM	Brown homogenous area Blue-gray peppering Follicular plugging Perifollicular gray ring
Case-3	Female/30	Bilateral ear, pre- and-post-auricular area	Two	SLE Lupus nephritis ISM	Brown and white homogenous area Brown and blue-gray peppering Peri-appendageal accentuation of peppering Follicular plugging Perifollicular gray ring Brown pigment network

SLE = Systemic lupus erythematosus.

with both benign and malignant pigment tumors such as flat seborrheic keratosis, large cell acanthoma, solar lentigo and lentigo maligna. Clinically, it may not always be possible to diagnose and differentiate pigmented macular CCLE from other causes of acquired hyperpigmentation, including post-inflammatory hyperpigmentation. Dermoscopy may help in solving this problem. Dermoscopic features described for various acquired facial pigmentary conditions in skin of color are as follows: melasma and PCD show a brown pigmentation, pseudo-pigment network, brown to gray dots and globules, and characteristically spare the peri-appendageal areas; LPP is found to have diffuse brown pigmentation, pseudo-pigment network, slate-gray to blue dots and globules, hem-like pigment pattern, and perifollicular and peri-eccrine brown to blue-gray dots. In the future, immunohistochemistry stain for melanin and /or melanocytes over a larger number of patients, as well as electron microscopy, may help to support dermatoscopy over the naked eye in the diagnosis of CCLE.

In conclusion, the clinical diagnosis of the pigmented macular variant of CCLE can be challenging due to its similarity to various facial pigmentary lesions. We describe the clinico-dermoscopic features in three patients with skin of color. Future studies will determine the role of dermoscopy in diagnosing and differentiating this rare entity from its clinical mimics.

References

1. George R, Mathai R, Kurian S. Cutaneous lupus erythematosus in India: immunofluorescence profile. *Int J Dermatol.* 1992;31(4):265-269. DOI: 10.1111/j.1365-4362.1992.tb03568.x. PMID: 1634292.
2. Boyd AS. Localized chronic cutaneous lupus erythematosus masquerading as pigmented lesions: a new clinical subset?. *Lupus.* 2006;15(5):292-295. DOI: 10.1191/0961203306lu2304xx. PMID: 16761504.
3. Krueger L, Saizan A, Stein JA, Elbuluk N. Dermoscopy of acquired pigmentary disorders: a comprehensive review. *Int J Dermatol.* 2022;61(1):7-19.DOI: 10.1111/ijd.15741. PMID: 34235719.
4. Chatterjee M, Neema S. Dermoscopy of Pigmentary Disorders in Brown Skin. *Dermatol Clin.* 2018;36(4):473-485. DOI: 10.1016/j.det.2018.05.014. PMID: 30201156.