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Café au Lait macules or Cause for Concern? Unravelling it's secret with histology and dermoscopic findings

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Abstract

Café au lait macules (CALMs) are common, benign, light-brown skin patches that usually appear in childhood. Typically under 2 cm in size and found on sun-protected skin, they are present in about 2–3% of newborns and a third of school-aged children. While single CALMs are harmless, multiple lesions may signal underlying disorders like neurofibromatosis type I (NF1) or McCune-Albright syndrome (MAS). A case study of a 3-year-old boy with enlarging CALMs highlights the need for clinical, dermoscopic, and histological assessments to rule out associated syndromes such as NF1, MAS, segmental neurofibromatosis, and Watson syndrome, though the child showed no signs of these. CALMs carry no risk of malignancy, but cosmetic concerns often lead to treatment. High-fluence Q-switched 1,064-nm Nd:YAG laser therapy has proven safe and effective, achieving over 75% clearance in about half of patients with minimal side effects, making it a valuable option for managing these lesions.

Keywords: Café au lait macules, dermoscopy, hyperpigmented lesions, laser therapy, pediatric age group.

Dear Editor,

Café au lait macules (CALMs) are common, benign, flat hyperpigmented lesions typically presenting at birth or in early childhood. These lesions are usually less than 2 cm in diameter and more frequently occur on sun-protected areas. While often isolated and clinically insignificant, multiple CALMs may serve as important diagnostic markers for systemic disorders such as neurofibromatosis type 1 (NF1) and McCune-Albright syndrome (MAS). Six or more CALMs are strongly suggestive of NF1, necessitating thorough evaluation. [1]

A 3-year-old male presenting with gradually progressive, dark, well-demarcated lesions over the chest, back, bilateral upper arms, forearms, and right thigh over one year. The patient had no history of pruritus, pain, ocular or auditory abnormalities, though flat feet were noted. (Figure-1) The lesions crossed the midline with additional isolated lesions on the thigh. Clinical evaluation, dermoscopy, and histopathology were employed for differential diagnosis.

Café au lait macules are common, flat, hyperpigmented lesions present at birth or early childhood. Clinical, dermoscopic, and histological evaluations help differentiate them from similar lesions like freckles, lentigines, congenital melanocytic nevi, Becker nevus, post-inflammatory hyperpigmentation, and urticaria pigmentosa, each with distinct distribution, size, sun-responsiveness, hair association, and histological features.[2]

Dermoscopy under polarized light demonstrated a fine, light-brown reticular network with diffuse homogeneous pigmentation and scattered blue-gray dots, characteristic of CALMs. Notably, features such as perifollicular hyperpigmentation, hypertrichosis, or globular patterns—common in congenital melanocytic nevi (CMN)—were absent. CALMs also lack the globular or homogeneous patterns seen in CMN and can resemble normal skin patterns in darker skin tones.[3] (Figure 2)

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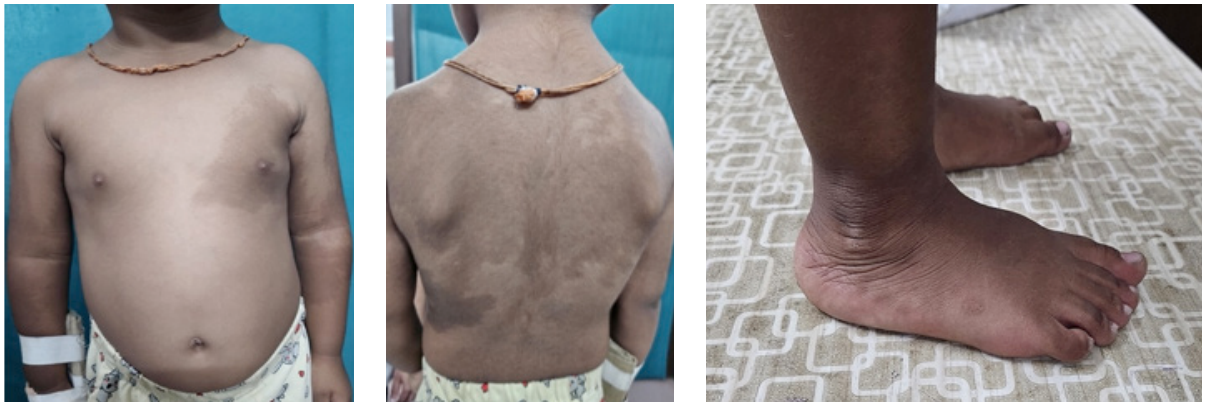

Figure 1a, 1b, 1c: Café au lait macules presenting as well dermarcated, hyperpigmented patch over left side of chest and bilateral arms, forearms with clubfoot

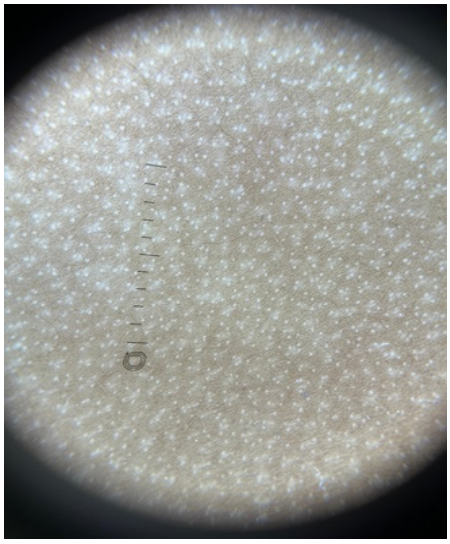

Figure-2: Dermoscopic image of café au lait macules (polarized, low power,10x), showing diffuse homogenous pigmentation

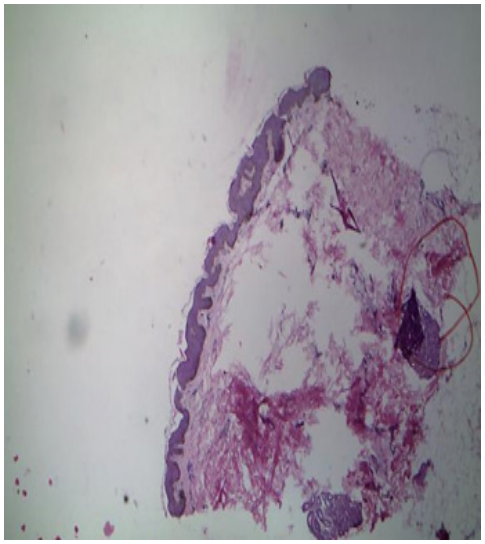

Figure-3: Mild epidermal hyperplasia with an increased number of evenly spaced, non-atypical melanocytes within the basal layer (10X)

Histopathological examination revealed mild epidermal hyperplasia with an increased number of evenly spaced, non-atypical melanocytes within the basal layer, without melanocyte nesting or dermal melanophages, confirming the diagnosis. (Figure-3)
There are many conditions associated with CALM such as- McCune-Albright syndrome, MAS- caused by postzygotic GNAS mutations leading to mosaicism, is characterized by CALMs, polyostotic fibrous dysplasia, and endocrine hyperfunction. CALMs in MAS typically have irregular "coast of Maine" borders, often involving the posterior neck, thorax, sacrum, and buttocks, with unilateral distribution. MAS manifests in the first decade with skeletal deformities (flat foot was noted in this case,Fig-4), fractures, and limb asymmetry, followed by endocrinopathies such as precocious puberty and hyperthyroidism.[1]
NF1 (von Recklinghausen's disease) involves multiple CALMs, axillary and inguinal freckling (Crowe's sign), neurofibromas, optic gliomas, Lisch nodules, and osseous abnormalities. Diagnosis requires two or more NIH-defined criteria, including six or more CALMs (>5 mm in prepubertal or >15 mm in postpubertal individuals), neurofibromas, plexiform neurofibromas, optic glioma, Lisch nodules, characteristic osseous lesions, or a positive first-degree family history.
Other differential considerations include segmental NF, confined to a single body segment without systemic involvement, and Watson syndrome, featuring CALMs, pulmonic stenosis, intellectual disability, and short stature. [4,5]
Management of CALMs is primarily cosmetic, as they carry no malignant potential. No medical therapy has proven effective. Laser therapy remains the treatment of choice. High-fluence Q-switched 1,064-nm Nd:YAG laser has demonstrated excellent results.[5]
This case underscores the importance of comprehensive dermatologic, dermoscopic, and histopathologic assessment in children presenting with multiple CALMs, while highlighting effective cosmetic treatment options.

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