Papulonodular Mucinosis in Childhood Subacute Cutaneous Lupus Erythematosus

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ABSTRACT

Papulonodular mucinosis in childhood subacute cutaneous lupus erythematosus (SCLE) is uncommon. We report a unique case of SCLE in a dark-skin individual. We describe a nine-year-old Nigerian boy who initially presented with recurrent, red, annular, discreet papules/plaques on scalp, face, upper arms, palms, lower abdomen, thighs and later with patchy alopecia, and subcutaneous nodules on both upper arms. Diagnosis was confirmed by histology, Alcian blue staining, presence of SSA antibody; and, patient responded to hydroxychloroquine and sun protective measures. This case highlights hyperpigmentation as a feature of SCLE in a dark-skinned individual and the involvement of SCLE in non-photosensitive body regions such as palms, thighs, and lower abdomen.

Keywords: papulonodular mucinosis, subacute cutaneous lupus erythematosus, Nigerian boy, hyperpigmentation, dark-skinned individual, non-photosensitive body regions

Mucinose Papulonodulaire dans le Lupus Érythémateux Cutané Subaigu de l'enfant

ABSTRAIT

La mucinose papulo-nodulaire dans le lupus érythémateux cutané subaigu de l'enfant (SCLE) est rare. Nous rapportons un cas unique de SCLE chez un individu à peau foncée. Nous décrivons un garçon Nigérian de neuf ans qui s'est initialement présenté avec des papules/plaques discrètes, rouges, annulaires et discrètes sur le cuir chevelu, le visage, les bras, les paumes, le bas-ventre, les cuisses et plus tard avec une alopécie inégale et des nodules sous-cutanés sur les deux bras. Le diagnostic a été confirmé par l'histologie, la coloration au bleu Alcian, la présence d'anticorps SSA; et, le patient a répondu à l'hydroxychloroquine et aux mesures de protection solaire. Ce cas met en évidence l'hyperpigmentation en tant que caractéristique du SCLE chez un individu à la peau foncée et l'implication du SCLE dans des régions corporelles non photosensibles telles que les paumes, les cuisses et le bas de l'abdomen.

Mots-clés: mucinose papulo-nodulaire, lupus érythémateux cutané subaigu, garçon Nigérian, hyperpigmentation, individu à peau foncée, regions du corps non photosensibles

Introduction

Papulonodular mucinosis (PNM) associated with lupus erythematosus (LE) belong to a heterogenous group of disorders called cutaneous mucinosis in which an abnormal amount of mucin is found in dermal layer of the skin.[1] There are two groups of cutaneous mucinosis: primary whereby mucin deposition is detected both clinically and histologically and secondary cutaneous mucinosis in which mucin deposition is an additional histological finding but is not detected clinically.[1] Lupus Erythematosus has been associated with both groups. Papulonodular Mucinosis, a primary cutaneous mucinosis has been reported largely in systemic LE. Only two cases of PNM in subacute cutaneous LE (SCLE) have been reported in the literature since PNM associated with LE was recognized as a disease entity in 1954 by Gold.[2] We report a case of PNM in a child with SCLE with its peculiar clinical features on the dark skin.

Case Report

A 9-year-old Nigerian boy presented in the last month of 2018 with multiple, red, recurrent, annular eruptions on scalp, face, trunk, upper and lower limbs of 5 years duration. Lesions were initially pruritic, scaly, and changed color from red to black with time.

Apart from the skin lesions, he was otherwise healthy. He had no known allergies and was not on any medication. His mother has a long history of discoid LE with scarring plaques on scalp and both upper arms. He is the second of two children. No similar lesions in his sibling.

Examination revealed multiple, reddish to hyperpigmented papules, plaques and patches on the scalp, face, earlobe, lower abdomen, upper arms, dorsum of hands and thighs. Some lesions coalesced into large annular, lichenified plaques with adherent scales (Figure 1).

There were old lesions on the scalp, pinna and upper limbs which healed as hyperpigmented macules and patches. There was no scarring, alopecia, or atrophy on any of the parts of the body involved. Plaques with adherent scales were observed on the interphalangeal folds of both hands while scaly dark/purple patches were seen on both palms (Figure 2). No nail abnormality detected.

Blood analysis showed a normal full blood count, liver, and renal function tests. Urine analysis did not show any proteinuria. Erythrocyte sedimentation rate was 39mm/hour (reference range 3-15mm/hour). Antinuclear antibody screen was positive, with a titer of 1:320. SS-A/anti-Ro was positive. Anti-double-stranded DNA and VDRL were not detected. Viral screening for Hepatitis B, C and HIV were non-reactive. Creatinine kinase and Anti-Jo 1 antibodies were negative. A punch biopsy from the upper arm revealed epidermal hyperplasia with prominent rete ridges, patchy vacuolar degeneration, melanin incontinence and deep perivascular and perifollicular lymphocytic infiltrate.

Patient was commenced on hydroxychloroquine 200mg twice daily and one-month course of oral prednisolone, lesions resolved within two months except for residual hyperpigmentation. Hydroxychloroquine was discontinued. Ten months later, patient presented with multiple asymptomatic subcutaneous nodules on lateral aspect of both arms and infiltrated plaques on occipital scalp with circular patches of non-scarring alopecia in same area that progressively increased in size and number. Biopsy taken from the plaque on the scalp and nodules on left upper limb showed marked intradermal deposits of mucin among collagen bundles (Figure 3b).

Hydroxychloroquine was recommenced and he was advised to strictly adhere to use of sun protection creams. He received two intralesional triamcinolone treatment. Presently, subcutaneous nodules and alopecia are resolving.

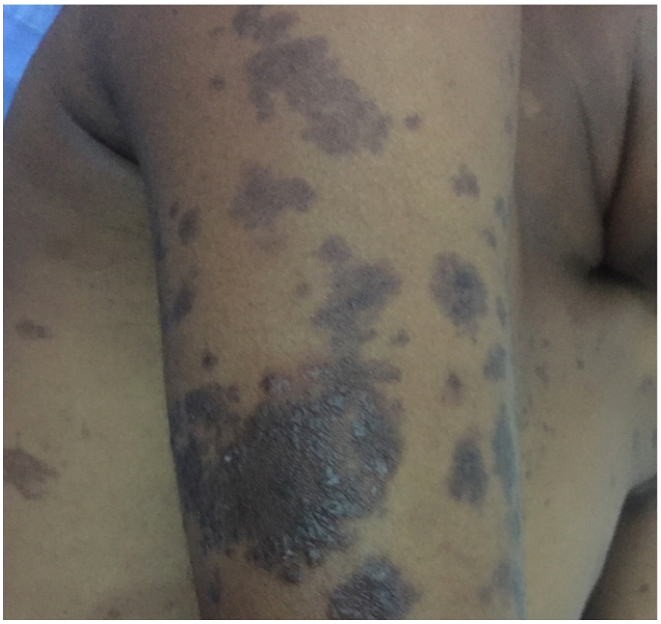

Figure 1: Multiple reddish to hyperpigmented, annular plaques and patches on lateral aspect of right upper arm

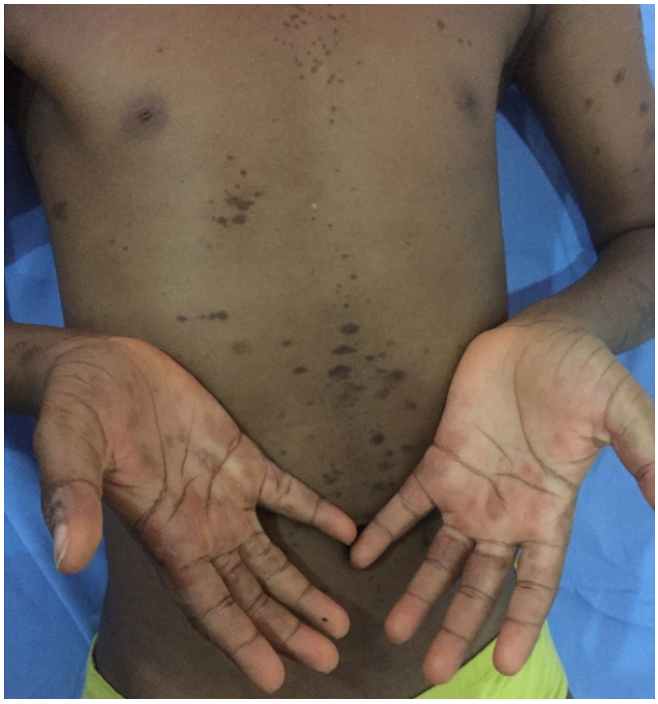

Figure 2: Dusky red scaly patches on both palms and hyperpigmented lesions on abdomen

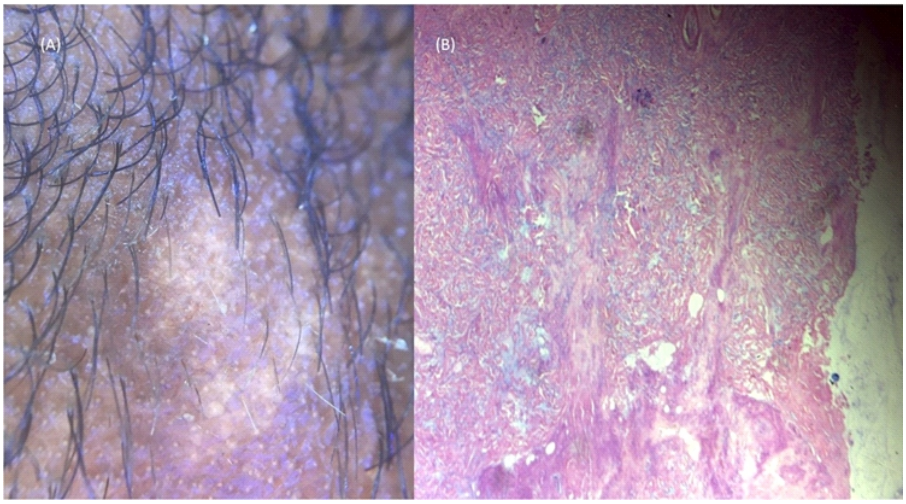

Figure 3: ***(A)*** picture of infiltrated nodular plaque and non-scarring alopecia; ***(B)*** Photomicrograph of alcian blue stain, original magnification x 10 showing mucin deposits in both papillary and reticular dermis

Discussion

Sontheimer et al's criteria for clinical diagnosis of SCLE include erythematous nonscarring papulosquamous/annular polycyclic and/or psoriasiform plaques characteristically distributed on face, neck, upper trunk, shoulders, extensor arms, dorsal hands and fingers with telangiectasia and hypopigmentation occurring at the center of the annular lesions.[3] SCLE is considered most prevalent among middle-aged Caucasian females; and its report in childhood and among other racial groups is uncommon. Literature search revealed 13 cases of childhood onset SCLE,[4-6] age ranging from 2 to 17 years including one case in a 2-year-old child of African American or Hispanic descent.[5] It is worthy of note that seven of this 13 cases were males. Although, our patient's lesions were the distinctive photosensitive SCLE pattern, he also had lesions on atypical regions like his lower trunk, palms, and thighs.

The post-inflammatory hyperpigmentation observed in our patient is not in tandem with the clinical features of SCLE reported in the literature is. Sontheimer et al.[7] classically described hypopigmentation of old lesions as a distinct feature of SCLE. Hypopigmentation have been attributed to melanocytic injury associated with the lichenoid tissue reaction seen in active SCLE.[7] Most of our dark skinned patients with lichenoid eruptions in Nigeria heal with hyperpigmentation and biopsy usually reveal melanin incontinence as seen in this case. Furthermore, this is the first report, to the best of our knowledge, of SCLE in an African patient. More reports in these group of patients are necessary to show the proper pattern of dyspigmentation that follow previously active SCLE lesions. So far, only two cases of PNM have been described in a child, both of whom had systemic LE[8-9] and none, to the best of our knowledge, have been described in a child with SCLE.

Exposure to sunlight have been implicated in the pathogenesis of PNM associated with LE.[2] Others have proposed that unknown cytokines or immunoglobulins stimulate normal fibroblasts to overproduce glycosaminoglycans.[1] Hormone is also a factor since PNM is observed more in males than females. Patient's mother has discoid LE and it is likely that these two cutaneous LE disorders could be linked genetically. Papulonodular mucinosis lesions are usually asymptomatic, skin colored and can be found within or outside typical LE lesions. Alopecia associated with PNM lesions in LE have been described only twice[10] in the literature. Alopecia is usually patchy, nonscarring and resembles alopecia areata but for the presence of papules/infiltrated plaques in the areas of alopecia as seen in our case.

Papulonodular mucinosis could predate, occur simultaneously, or appear years after LE. In the first report of PNM in SCLE[2], nodules were observed years after the first signs of SCLE. While in our case,

they occurred a few months after the clinical signs of SCLE have resolved and patient had discontinued hydroxychloroquine. Out of the various modalities employed in the treatment of PNM such as corticosteroids (topical, intralesional and systemic), antimalarial agents, methotrexate, excision, abrasion and laser; antimalarial agents and sun protection have been shown to be more effective, as seen in our patient. Thus, it may be safe to conclude that the course of PNM in SCLE is possibly related to the response of SCLE to treatment and to speculate that PNM that develops after resolution of SCLE may be an indication of the presence of subclinical SCLE.

Key Messages: Subacute cutaneous lupus erythematosus lesions can present as hyperpigmented macules and patches in dark-skinned individuals and lesions can also be found on non-photosensitive regions of the body. Intradermal mucinosis on scalp can cause non-scarring alopecia.

No conflict of interest to declare

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Written Informed Consent statement: The authors certify that written informed consent was obtained from the patient and his patients to publish this report in accordance with the journal's patient consent policy.

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