

A Case of Generalized Pruritic Lichen Nitidus with Palmoplantar Involvement - Pediatric Generalized Lichen Nitidus

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Abstract

Case Report

Lichen nitidus (LN) is a rare cutaneous disorder characterized by the presence of multiple, usually asymptomatic, small papules, most commonly localized on the neck, chest, forearms, abdomen and genitalia. Rare cases of generalized LN and of palmoplantar LN have been reported, and to our knowledge, no cases of concomitant generalized and palmoplantar LN have been described. We present the case of a 7-year old girl with generalized and palmoplantar pruritic LN. This cutaneous disorder has a pathognomonic histopathology characterized by a dermal lymphohistiocytic infiltrate in a ball in claw configuration. It is usually self-limited, but diffuse or persistent disease is indication for treatment.

Keywords: Lichen nitidus, generalized, pruritus, pediatric.

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INTRODUCTION

Lichen nitidus (LN) is a rare cutaneous disorder characterized by the presence of multiple small papules most commonly localized on the neck, chest, forearms, abdomen and genitalia. Rare cases of generalized LN and of palmoplantar LN have been reported, and to our knowledge, no cases of concomitant generalized and palmoplantar LN have been described. We present the case of a 7-year old girl with generalized and palmoplantar pruritic LN.

CASE REPORT

A 7-year old girl presented to our dermatology department for a diffuse pruritic papular eruption that started abruptly 4 months before her visit. Her past medical history was negative for diseases or any recent medication use. She was already treated with moisturizing and emollient creams, and antihistamines, without any improvement.

The dermatological examination showed disseminated small skin-colored papules over her face, neck, chest, abdomen, back, both arms and legs, with brownish hyperkeratotic papules and plaques on the palms, the lateral aspect of the fingers and soles. The presence of follicular pustules over her abdomen, and multiple impetiginized lesions on her scalp, face, and arms was also noted (*fig 1-3*).



Fig-1: Clinical view of palmar lesions



Fig-2: Clinical view of papular lesions on the thighs



Fig-3: Clinical appearance of multiple papular lesions on the face

No lesions on the mucous membranes, or nail dystrophy were present. There was no family history of a similar condition. The patient received oral ivermectin 1 tablet once per week for 3 weeks - according to her weight - for the treatment of possible scabies, but no improvement was noted. She was also given oral amoxicillin-clavulanate for 2 weeks with resolution of the impetiginized lesions. Blood tests including a complete blood count, renal and liver function tests, CRP, and anti-streptolysin-O (ASO), showed no abnormalities.

Therefore, a skin biopsy was done, and it showed: irregular acanthosis underlying a thin epidermis with areas of focal atrophy and marked well-circumscribed dermal lichenoid infiltrate. These changes were accompanied by variable parakeratosis, focal erosion of the epidermis and mild formation of

granulation tissue. The lichenoid infiltrate was mainly composed of mononuclear cells of the granulomatous type with some giant multinucleated cells with pigment incontinence (fig. 4, 5).

Special stains didn't reveal any additional histologic features.

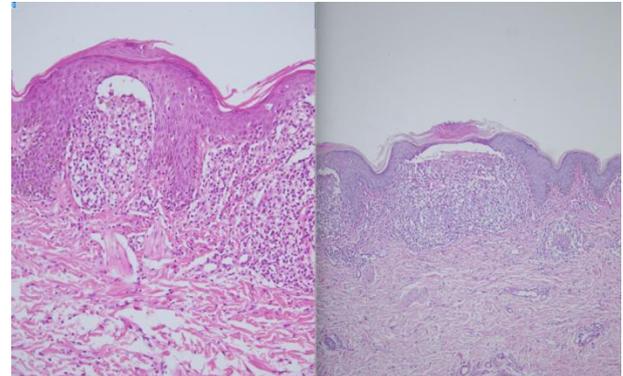


Fig-4,5: Histopathological appearance of a well-circumscribed lymphohistiocytic infiltrates

Based on the clinical and histologic findings, the diagnosis of generalized lichen nitidus is made. The patient received a treatment based on topical corticosteroids.

DISCUSSION

Lichen nitidus (LN) is a rare idiopathic chronic cutaneous disorder that affects most frequently children and young adults. It is typically characterized by multiple, discrete, skin-colored, shiny round papules, 1 to 2 mm in diameter, that are predominantly localized over the neck, chest, abdomen, arms, forearms, and genitalia, and rarely on the nails, palms and soles, and mucous membranes. The lesions are usually asymptomatic, but can sometimes be mildly pruritic. [1] Koebner phenomenon is also usually present.

Several uncommon variants of LN have been described, including confluent, vesicular, hemorrhagic, palmar and plantar, follicular, perforating, purpuric, linear and generalized forms, [2,3] all of which share a pathognomonic histology: atrophic epidermis, a well circumscribed lymphohistiocytic infiltrate in the papillary dermis, and elongated rete ridges in a ball in claw or ball in clutch configuration [4].

Generalized LN is characterized by the widespread distribution of the papular lesions over the body. Our patient presented with the universal type where the lesions are over nearly the entire body surface, and this subtype of LN is particularly rare [5].

Palmoplantar LN can be diagnosed by the presence of similar papules on the palms and soles and rarely by diffuse palmoplantar hyperkeratosis. Hyperkeratotic plaques with pits are considered suggestive for the diagnosis [6,7].

The exact etiology of LN is unknown, but familial occurrence has been reported, suggesting a possible genetic predisposition, which could possibly render individuals susceptible to some environmental factors that induce the eruption [8] Generalized LN has also been reported in association with genetic disorders such as Down syndrome [9, 10] and Russel-Silver syndrome [11]. In our case, family history and physical examination were negative for any genetic or other related disorders.

LN is usually a self-limited cutaneous disorder, which resolves without sequelae, but with an unpredictable evolution. Hyperpigmentation after resolution has been reported [12]. Medical treatment should be considered if the disease is persistent, diffuse, or causing pruritus or cosmetic discomfort in the patient [13].

Several treatments including topical and systemic steroids, phototherapy (narrow UVB band) [14], photochemotherapy, selective H1 antagonist, itraconazole, isoniazid, and isotretinoin [13,14], topical calcineurin inhibitors (pimecrolimus 0.1%) [15], have been tried. Since our patient had a diffuse pruritic disease, she was given topical steroids with close follow-up.

CONCLUSION

Lichen nitidus (LN) is a rare cutaneous disorder characterized by the presence of multiple small papules most commonly localized on the neck, chest, forearms, abdomen and genitalia. Rare cases of generalized LN and of palmoplantar LN have been reported, and to our knowledge, no cases of concomitant generalized, and palmoplantar LN have been described. This diagnosis should be kept in mind in the differential diagnosis of a pruritic skin eruption in childhood.

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