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Case Report

Unilateral prurigo nodularis: a rare presentation

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ABSTRACT

Prurigo Nodularis (PN) is a rare chronic skin disorder of unknown etiology. Here we are describing a case of 14 year old girl having prurigo nodularis with no other systemic illness.

Keywords: Prurigo nodularis, Unilateral, Histopathology

INTRODUCTION

Prurigo nodularis was first described by Hyde et al.[1] in 1909 as pruritic nodules on the extensor surfaces of the lower extremities in middle-aged women. Prurigo nodularis usually seen as multiple, intensely pruritic, excoriated nodules erupting on the extensor surfaces of the limbs secondary to itching or rubbing. Prurigo nodularis is still a condition of unknown etiology. Here we are describing a case with unilateral prurigo nodularis with no other systemic illness.

CASE REPORT

A 14 year old girl presented with complaints of gradually progressive, multiple, raised, hyperpigmented lesions present only on the left lower limb associated with itching for last 1 year. She had taken multiple treatment (oral and topical) with partial relief in the symptoms.

There was no history of oral soreness, insect bites or of significant weight loss. No history suggestive of atopy or any drug intake prior to appearance of the lesions. General and systemic examinations were essentially normal. On mucocutaneous examination, there were multiple, hyperpigmented, hyperkeratotic scaly papules and nodules varying from 0.75 x 0.75 cm to 2 x 2 cm in size present in a linear arrangement on the left lower limb extending from mid-thigh to the dorsum of foot, mainly over the extensor aspect. An interesting finding was the presence of hypertrichosis over the affected limb, but no significant relevance could be found. There were no similar lesions on any other sites. Mucosae, hair and nails were normal.

Differential diagnoses made were: hypertrophic lichen planus, prurigo nodularis, lichen striatus / adult blaschkitis, pemphigoid nodularis, nodular scabies.

Routine laboratory work-up was normal. On histopathologic examination, there was sparse superficial perivascular lymphocytic infiltrate with mild to moderate irregular epidermal hyperplasia. The papillary dermis was thickened with papillomatosis and showed thickened bundles of collagen in vertical array. The capillaries in papillary dermis were increased in number and were thick walled. The granular layer was thickened and the stratum corneum showed marked compact orthokeratosis.

The diagnosis of unilateral prurigo nodularis was made on the basis of history, examination and histopatholgy report.

We advised measures to reduce excoriation including cutting the nails very short, wearing gloves at night and occlusion of the involved regions of skin with bandages. Potent topical steroids along with oral antihistamines were given. Patient was lost to follow up therefore no further study was undertaken.

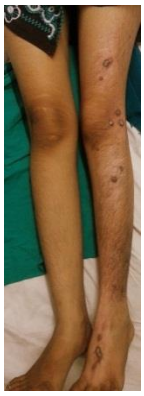

Figure 1: Hyperpigmented hyperkeratotic scaly popular and nodular lesions on the extensor surface of the leg.

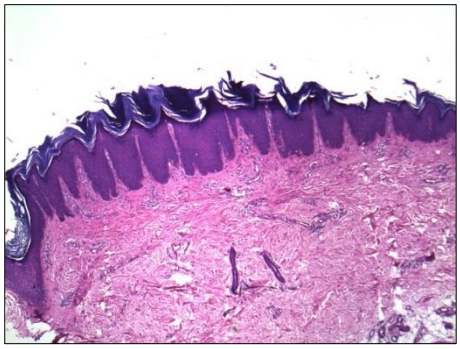

Figure 2(a): H&E (10x) section showing epidermal hyperplasia with papillamatosis, and papillary dermis showing vertical array of thickened collagen.

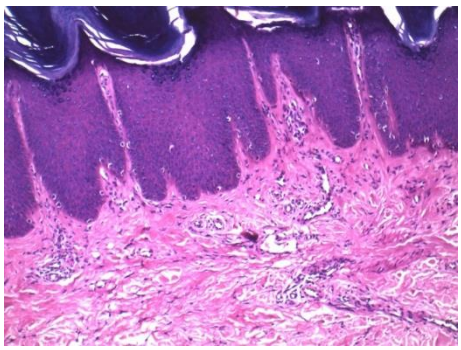

Figure 2(b): H&E (40x) section showed marked compact orthokeratosis, perivascular lymphocytic infiltrate and bundles of collagen.

DISCUSSION

Prurigo Nodularis (PN) is a chronic, highly pruritic condition characterized by the presence of hyperkeratotic, excoriated, pruritic papules and nodules, with a tendency to symmetrical distribution. Lesions are usually grouped and numerous but may vary in number from 2-200.[2] The disease is relatively rare and can occur at any age, but it is more commonly reported in middle-aged women.

PN is one of the most challenging of all chronic skin disorders in terms of establishing its aetiology and determining treatment strategies.[3,4] These issues are related to what triggers it, as an important first step in therapy is to identify the underlying cause and treat the condition accordingly.

Various factors have been identified to cause PN, such as insect bites, folliculitis and nummular eczema, mycobacterial infection, biliary obstructive disease (intrinsic, extrinsic or drug), hepatitis C, chronic kidney disease, Hodgkin's disease, leukemia, anemia, polycythemia vera, venous stasis, solid tumors, carcinoid syndrome, hypothyroidism and hyperthyroidism, diabetes, parasitic diseases, drug reactions, gluten induced enteropathy or other malabsorption.[5-10]

The differential diagnosis should be considered with the hypertrophic lichen planus, prurigo nodularis, lichen striatus / adult blaschkitis, pemphigoid nodularis, nodular scabies because of overlapping clinical symptoms. PN has distinct histopathological pattern that differentiate it from other diseases.

The PN is quite resistant to conventional therapies, so treatment modalities are limited and has unsatisfactory results. Antipruritic agents, antihistamines, corticosteroids, UV light,[11] cryotherapy,[12] vitamin D_3,[13] capsaicin,[14] cyclosporine,[15] thalidomide[16] and naltrexone have been used to treat PN.

In conclusion; unilateral PN is a rare disorder with unknown aetiology. Histopathological examination should always be carried out to make correct diagnosis.

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