Juvenile pemphigoid nodularis: Report of a rare case

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ABSTRACT

Bullous pemphigoid is an autoimmune blistering disease that is rare in childhood. Pemphigoid nodularis is a variant of BP that is exceedingly rare in children. Pemphigoid nodularis is characterized by overlapping clinical features of both prurigo nodularis and BP. We report here a case of pemphigoid nodularis in an 11-year-old boy.

Key words: Bullous pemphigoid, juvenile, pemphigoid nodularis

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INTRODUCTION

Bullous pemphigoid (BP) is the most common autoimmune subepidermal blistering disease. The disease typically presents in the elderly, with onset after 60 years of age. The disease also occurs but rarely in children. There is no known ethnic, racial, or gender predilection. Skin lesions are characterized by tense blisters arising on a normal or erythematous base, most commonly over lower abdomen, inner thighs, and flexor surfaces of limbs. Mucous membrane involvement may be seen in approximately 10-35% patients and is mostly limited to oral mucosa. The uncommon variants of BP are urticarial plaque and eczematous lesion, prurigo nodularis-like, vegetating lesion, localized and dyshidrosiform type. Pemphigoid nodularis is a rare variant of BP showing features of both nodular prurigo and BP. The condition appears to be more common in females than males. A few cases of pemphigoid nodularis have been reported earlier. Most of the patients belong to middle age and older age groups. This is the youngest patient of pemphigoid nodularis and also the first case to be reported from the eastern part of India.

CASE REPORT

An 11-year-old boy, born of nonconsanguineous marriage with normal developmental milestones, presented to us with extremely pruritic papulonodular lesions over bilateral shins, dorsum of feet, and nape of the neck for a duration of 5 years. The lesions first appeared over the shins and then on the dorsum of the feet. Similar lesions also appeared over the nape of the neck about 1 year earlier. There was no past history of similar illness or any other significant medical or surgical history. There was no history of trauma-induced blister, drug intake prior to the onset of blisters, and no history of photosensitivity was obtained. Eruption and formation of teeth were normal. The patient was being treated with various topical medications, details of which could not be elicited. No family history of similar illness was obtained.

Clinical examination revealed multiple, symmetric, skin-colored and hyperpigmented papules and nodules involving both shins and dorsum of feet [Figures 1 and 2]. On the nape of the neck lesions were slightly erythematous papules with excoriations [Figure 3]. A few nodules were seen to be surmounted by tiny vesicles [Figure 4]. Other parts of the skin, mucosae, hair, and nails revealed no abnormality. There was no regional lymphadenopathy. Tzanck smear was negative for acantholytic cells. Routine laboratory tests, including serum biochemistry, were normal.

Histopathological examination showed hyperkeratosis, subepidermal cleft with neutrophilic infiltration within the upper dermis [Figure 4]. The subcutis was normal.

Our differential diagnoses were epidermolysis bullosa pruriginosa, prurigo nodularis, and nodular pemphigoid. Direct immunofluorescence (DIF)

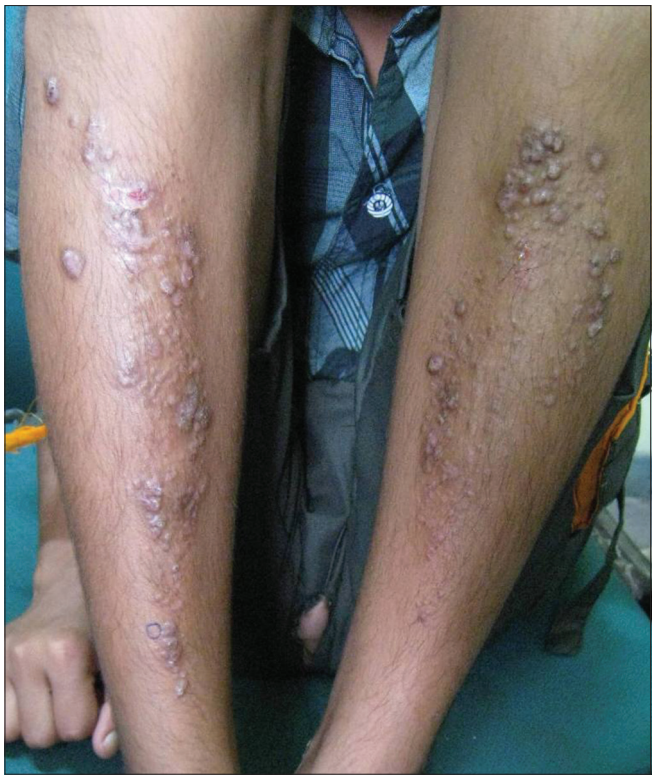

Figure 1: Skin-colored and hyperpigmented papules and nodules over bilateral shins

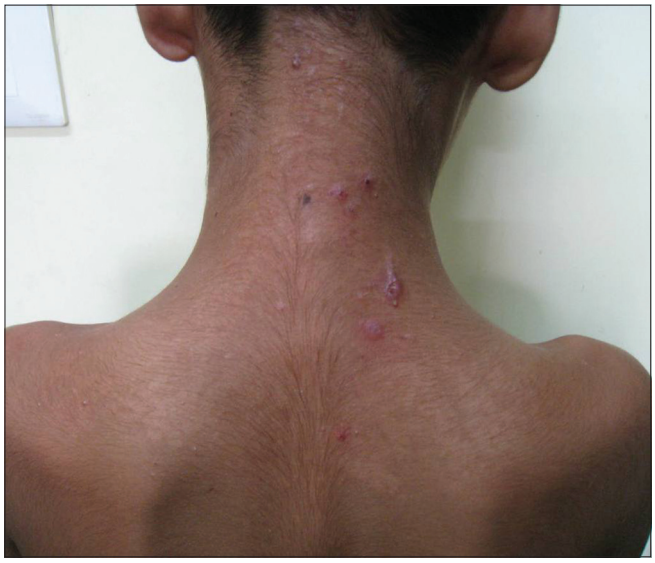

Figure 3: Erythematous excoriated papules over nape of the neck

testing showed linear deposition of IgG and IgM along the dermoepidermal junction, corroborating the diagnosis of pemphigoid nodularis, a variant of BP.

Our patient responded well to prednisolone. He was prescribed oral prednisolone starting at 20 mg once daily and the dose was tapered off over a three-month period to 5 mg once daily. By this time pruritus decreased and lesions also decreased in size and number.

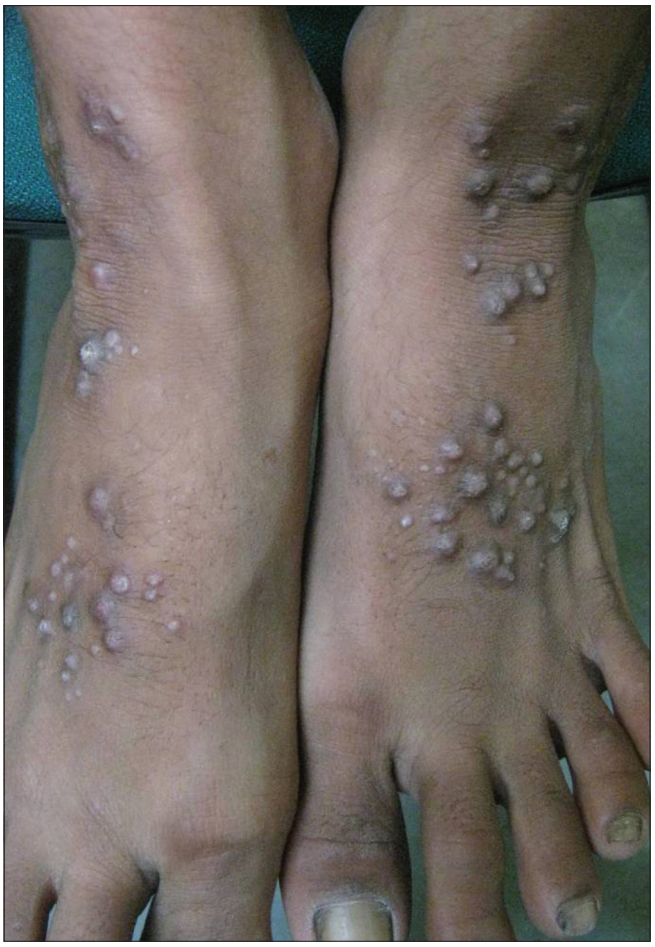

Figure 2: Hyperpigmented papules and nodules over dorsum of feet

DISCUSSION

Pemphigoid diseases were first differentiated from pemphigus in 1953 by Lever. A decade later, Jordon and colleagues showed that patients with BP had tissue-bound and circulating autoantibodies directed against the dermoepidermal junction.[1]

Apart from the classic bullous type, several other clinical variants of BP have been described such as localized, nodular, vegetating, childhood, erythrodermic, lichen planus pemphigoides, and drug-induced forms.

Autoantibodies in BP are directed against two hemidesmosomal proteins, designated BP180 and BP230. BP230 localizes intracellularly and is associated with the hemidesmosomal plaque, whereas BP180 is a transmembrane glycoprotein with an extracellular domain consisting of approximately 1000 amino acids. In the majority of BP sera, circulating antibodies to BP180 are detected, and their serum levels correlate with disease activity.[2]

Pemphigoid nodularis is one of the rarer variants of BP characterized by overlapping clinical features of both

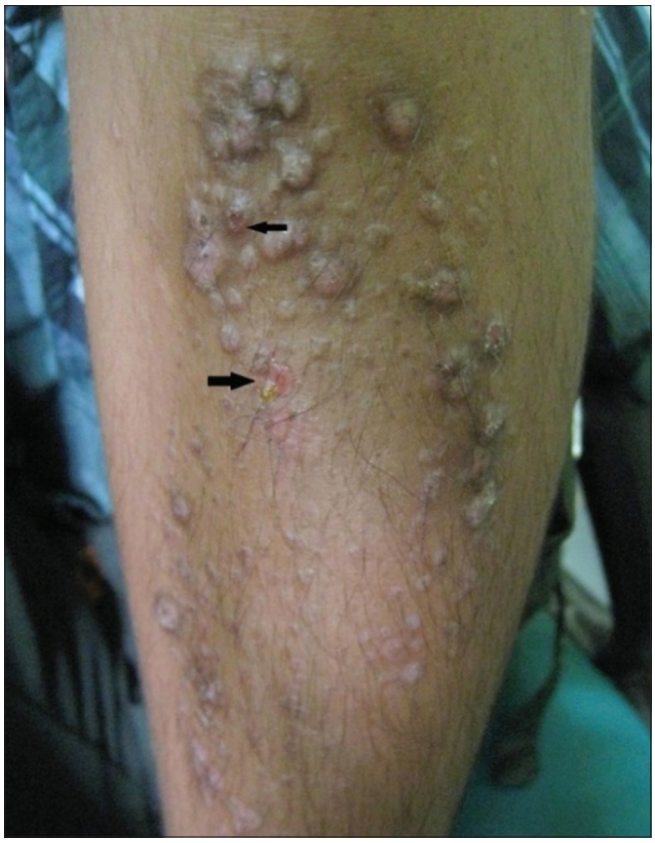

Figure 4: Few nodules over shin surmounted by tiny vesicles

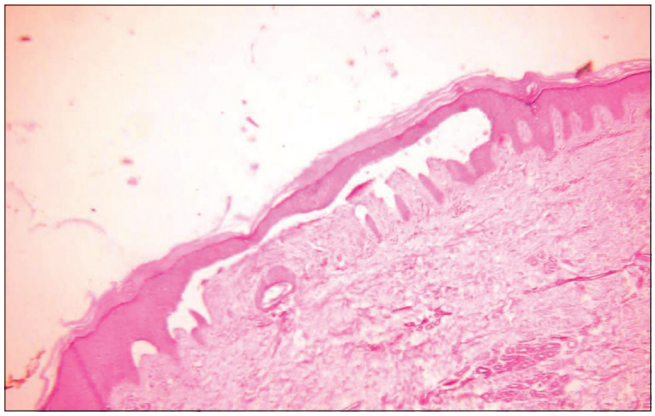

Figure 6: Histopathology of skin lesion showing hyperkeratosis, subepidermal cleft with chronic inflammatory infiltrate in the upper dermis. H and E, ×10

prurigo nodularis and BP. Blisters may precede the development of nodules[3-5] or may develop during the course of the disease.

The pathogenesis of this condition is not known properly. One hypothesis is that the nodules develop as a consequence of persistent scratching in predisposed individuals who have subclinical BP, which may subsequently be manifested clinically.

In pemphigoid nodularis, the immunopathological findings are identical to those of BP.[6] Autoantibodies in pemphigoid

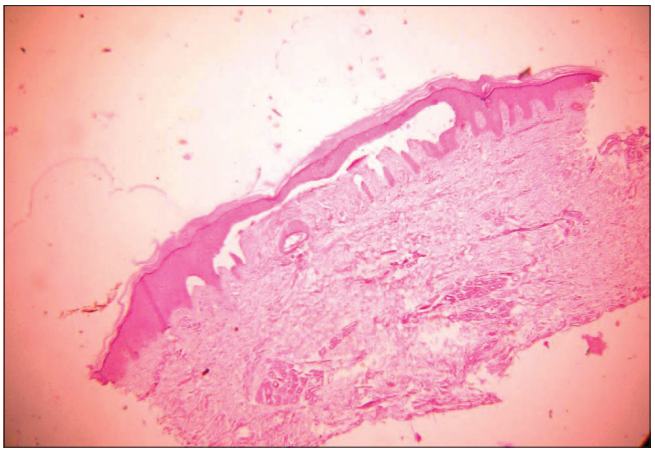

Figure 5: Histopathology of skin lesion showing hyperkeratosis, subepidermal cleft. H and E, ×4

nodularis target the same epitope within the NC16A domain of BP180 as autoantibodies in BP of adulthood.[7]

Provost *et al*.[8] first described BP in two female patients presenting with nodular prurigo-like lesions over the arms and legs. Both direct and indirect immunofluorescence studies were diagnostic of BP.

The term "pemphigoid nodularis" was coined by Yung *et al*.,[9] in 1981. They described a female patient of BP who, many years later, developed pruritic nodular lesions and subsequently diagnosed as pemphigoid nodularis.

Most of the patients of pemphigoid nodularis reported till date belonged to older age group.[4,5,9,10] Provost *et al*.[8] reported a 24-year-old woman with pemphigoid nodularis. A single case of juvenile pemphigoid nodularis has been reported by Ratnavel *et al*. who described a 15-year-old male presenting with bullous lesions and subsequently developed pruritic nodular lesions.

The histological and immunological features of pemphigoid nodularis are those of classic BP, with added epidermal hyperkeratosis suggesting that pemphigoid nodularis is a variant of BP.

Differential diagnoses considered in our case included epidermolysis bullosa pruriginosa and prurigo nodularis. Histopathology and DIF findings could exclude the two conditions.

Treatment of pemphigoid nodularis is difficult. Usually, it does not respond to topical corticosteroid alone. Most of the patients require systemic immunosuppressive therapy with prednisolone, dapsone and azathioprine, cyclophosphamide either alone or in combination. The disease runs a chronic course and is often resistant to treatment.

Although pemphigoid nodularis predominantly affects elderly women, it may present in younger age group also. Here we describe the youngest patient of pemphigoid nodularis reported till date.

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