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CASE REPORTS

BULLOUS PEMPHIGOID IN CHILDHOOD

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A young child with clinical presentation and histology like chronic bullous dermatosis of childhood and immunopathology of bullous pemphigoid is being reported to make the readers aware of its existence in Indian population and to stress the importance of immunopathology in bullous disorders of childhood.

Key Words : Childhood bullous pemphigoid, Immunopathology

Introduction

Bullous pemphigoid (BP) is an autoimmune bullous disorder mostly encountered in the elderly. It is uncommon in children and less than 50 cases have been reported.[1] Clinical presentation of BP is similar in both adults and children but for more frequent involvement of the oral mucous membrane in children.[2] Almost all patients show deposition of IgG and/or C_3 at the basement membrane zone (BMZ) on direct immunofluorescence (DIF). Indirect immunofluorescence (IIF) is positive for IgG at BMZ in 70% of adult cases.[3] Clinically it is often confused with chronic bullous dermatosis of childhood and it is very difficult to confirm the diagnosis on histology unless an immunofluorescent or immunohistochemical examination is carried out.

Case Report

A 5-year-old boy was referred for a persistent, severely pruritic, urticarial rash on the trunk, face, perineal area and proximal limbs associated with occasional blistering. He was not responding to antihistamines and relapsing on withdrawl of steroids. On examination multiple, urticarial plaques were present on the lower back, buttocks, perineal area, groins, thighs, upper arms and trunk (Fig. 1). Mucosal surfaces were

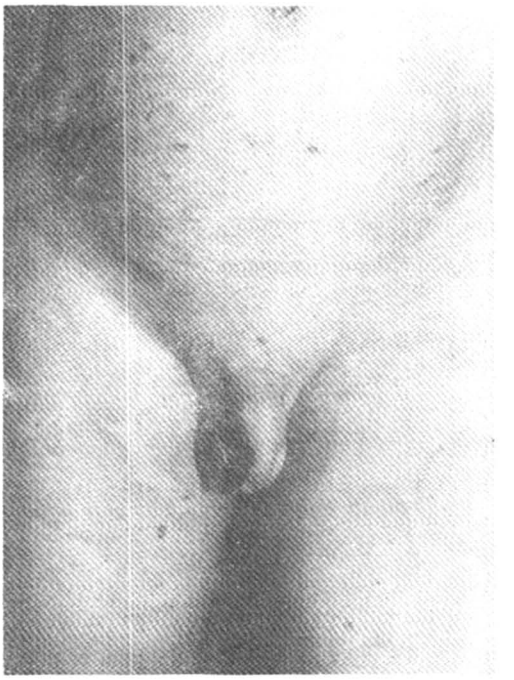

Fig. 1. Urticarial plaques on the abdomen, groins and pubic area.

free of lesions and so were the palms and soles. Skin biopsy was taken and submitted to histopathology and immunohistochemical staining. On the third day the child was seen again due to appearance of multiple vesicles, bullae and crusts. Most of them were on

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urticarial plaques, at places 'string of pearl' appearance was present. The same treatment was continued with a second possibility of bullous pemphigoid. Total and differential leucocyte counts revealed a normal total count with eosinophilia (13%). Haemoglobin estimation, G-6PD estimation, liver and renal function tests were within normal limits. The histology showed a subepidermal bullae with fibrin and a few eosinophils and polymorphs. There was moderate granulocytic infiltrate in the upper dermis with multiple eosinophils (Fig. 2). Direct immunohistochemical examination of the perilesional skin revealed deposits of IgG at the BMZ in a linear fashion (Avidin-Biotin Complex method) (Fig. 3).

He was weaned off steroids and

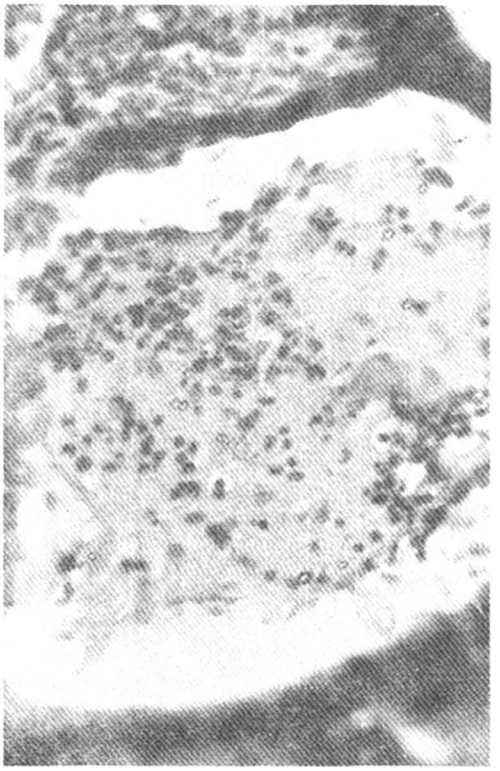

Fig. 2. Multiple eosinophils and polymorphs in the upper dermis (H & E x140).

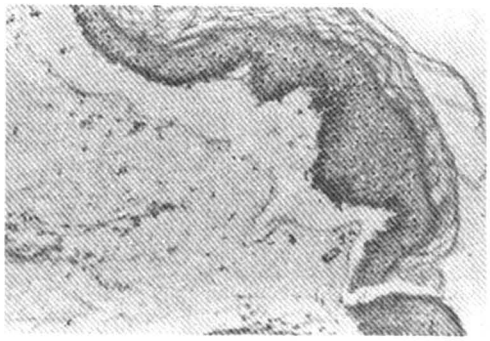

Fig. 3. IgG deposits at the BMZ (ABC-immunoperoxidase stain).

dapsone 75 mg per day was begun keeping in mind the possibility of chronic bullous dermatosis of childhood due to periorificial localization of the lesions. The lesions resolved with dapsone alone on the 4th day and no new lesions appeared 7th day onwards. The dose was reduced to 50 mg/day after 2 weeks of remission and the child is now free of lesions except for a short period when dapsone was stopped by his parents for 2 weeks. Further the dose was reduced to 25 mg of dapsone daily for 4 weeks and now he is on 25 mg of dapsone alternate days.

Discussion

It was Lever[4] who first differentiated BP from pemphigus vulgaris on basis of morphology and histopathology. Kim and Winklemann[5] further differentiated BP in childhood from dermatitis herpetiformis of childhood. Before the application of immunopathologic techniques to clinical dermatology earlier cases of childhood BP were probably categorised as dermatitis herpetiformis of childhood.[6] Bean et al[7] described the first case of childhood BP with immunofluorescence findings.

The diagnostic crtieria used to diagnose childhood BP so far in the literature[1] are: (1)

patients 18 years of age or younger with the clinical appearance of tense bullae on erythematous or non-erythematous skin with or without mucous membrane involvement, and routine histopathologic study showing subepidermal bulla formation with a variable amount of eosinophils; and more importantly (2) DIF showing linear deposition of IgG and/or C_3 as the major immunoreactant(s) at the BMZ and/or a positive IIF showing IgG antibodies reactive with antigen(s) at the BMZ.

The clinical presentation is similar to adults, facial involvement is common in children and can be used as an additional diagnostic clue.[2] Immunopathologic studies of the skin and serum are the most important diagnostic tools to differentiate childhood BP from other acquired immunobullous dermatosis. Direct IF staining of perilesional skin shows linear IgG deposits along BMZ in all patients; complement C_3 is found in all skin lesions and at times may be the only immunoreactant present. Other immunoreactants such as IgA, IgM and IgE may also be found rarely. Indirect immunofluorescence testing of the serum shows IgG antibodies to BMZ in 70% patients.[3]

Treatment of choice for BP is prednisone in a dosage of 1-2 mg/day but there are encouraging reports of response to sulfones alone.[8] We gave dapsone considering the relapse on withdrawl of oral steroids. He responded very well and has had no recurrence apart from a short period when the parents stopped dapsone for 2 weeks. The course of childhood BP is protracted but self limited. With therapy childhood BP usually follows a benign course. The majority of cases enter remission within 1 year. Overall childhood BP has an excellent prognosis.

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