ISSN: 2581 – 3633
PubMed - National Library of Medicine - ID: 101745081

International Journal of Medical Science and Advanced Clinical Research (IJMACR)

Available Online at:www.ijmacr.com

Volume – 6, Issue – 1, Janaury - 2023, Page No. : 652 - 658

Varying clinical presentations of childhood pemphigus vulgaris - A case report

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How to citation this article: Dr. Sreevidya Suresh M, Dr. Patnala Guru Prasad,Dr. Kethireddi Susmitha Divya,Dr. Karri Sudhir Babu,"Varying clinical presentations of childhood pemphigus vulgaris - A case report", IJMACR-January - 2023, Volume – 6, Issue - 1, P. No. 652 – 658.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Childhood Pemphigus vulgaris is a rare paediatric vesiculobullous disorder. It presents with flaccid blisters and mucosal involvement similar to adult pemphigus vulgaris. We report a case of childhood pemphigus vulgaris in a 10-year-old boy with 3 different clinical presentations within a period of 1 year. Initial presentation was similar to toxic epidermal necrolysis with widespread skin and mucosal involvement. Subsequently child presented with limited disease with few discrete tense vesicles. Both these initial presentations had diagnostic difficulty as the clinical picture was similar to other childhood bullous disorders. Finally, the child developed multiple flaccid vesicles along with mucosal involvement. Histopathology and immunofluorescence confirmed the diagnosis of Pemphigus vulgaris. Child was treated successfully with prednisolone and dapsone.

Keywords: Pemphigus Vulgaris, Child, vesiculobullous disorder

Introduction

Childhood pemphigus vulgaris constitutes only a small proportion of the vesiculobullous disorders of childhood.[1,2] Clinical presentation can be similar to as that of the adult, with flaccid blisters that produce erosions after rupture, painful oral and genital erosions.[1,3,8] Auto antibodies are directed primarily against the desmoglein-3 with or without involvement of desmoglein-1. These auto antibodies are responsible for the loss of cohesion between the epidermal cells and subsequent blister formation.[1] Histological and immunological features are similar to adult pemphigus vulgaris.[1,2,8]

Early diagnosis is of paramount importance as childhood Pemphigus Vulgaris can be managed well with early initiation of treatment.

Case report

A 10-year-old boy presented with extensive erosions all over the body with genital and oral erosions of 10 days duration (Fig 1-3). Patient was febrile, and nikolsky was positive. Histopathological findings were nonspecific. A diagnosis of toxic epidermal necrolysis was kept and patient was started on with systemic steroids, intravenous immunoglobulin and cyclosporine. All the lesions were resolved and the patient got discharged after 1 month of hospital stay. 5 months later the boy developed painless oral and scrotal erosions followed by few discrete tense vesicles containing clear fluid over the forehead along with crusted papules (Fig 4-6). Nikolsky was negative. Tzank smear showed multiple acantholytic cells. All other hematological parameters were within in normal limits.

Biopsy and immunofluorescenceshowed a suprabasal blister and intercellular deposits IgG and c3 confirming the diagnosis of pemphigus vulgaris (Fig 7,8).

Patient was started on with prednisolone 1mg /kg/day and dapsone 1mg/kg/day, oral erythromycin and other supportive measures. All the skin lesions were healed and the patient was discharged with dapsone 1mg/kg on the 15th day of admission. The patient was under regular follow up for a period of 1month and was kept on dapsone 0.5 mg/kg dose daily. 3 months later child developed multiple vesicles over the body along with oral erosions (Fig 9,10). The number of vesicles were more as compared to the prior episode. Parents revealed that they stopped taking all medications since 2 months which resulted in relapse. He was restarted on with dapsone 1mg/kg/day and prednisolone 1mg/kg/day and all the lesions were subsided within 2 weeks. He is under regular follow up and is on dapsone 0.5 mg/kg/day.

Discussion

Childhood pemphigus vulgaris constitutes 1.4 – 3.7 % of all pemphigus vulgaris cases.[3] Pemphigus in children younger than 12 years is known as childhood pemphigus vulgaris and in those between 12 to 18 years of age are called Juvenile pemphigus.[3,4]Drugs, herpes virus infection, bacterial infection and malignancies are considered as some of the triggering factors.[3,5-7]High incidence of genital and ocular involvement distinguishes the childhood variant from adult Pemphigus vulgaris.[3,9] Oral lesions are the initial presentation in most of the cases.[1] Our case initially developed oral and genital erosions followed by extensive skin lesions. This was followed by limited disease with few discrete tense vesicles after 5 months. The patient presented with a classic pemphigus vulgaris clinical picture later after 3 months.

The differential diagnosis of PV in children includes entities affecting mucocutaneous tissues or predomonantly the oral mucosa or some affecting predominantly the genitalia (Table 1).[3]

Histopathological findings of Suprabasalclefting with row of tombstone appearance and intercellular positivity for IgG and C3 in a fishnet pattern in direct immunofluorescence confirms the diagnosis.

Systemic corticosteroids remains the drug of choice. Higher doses of prednisolone 1mg/kg per day should be administered from the beginning which can be slowly tapered according to the clinical response.[3,7,10,11] The frequency of side effects of steroids is higher in children due to the growth and development in childhood.[3,7,9,13,] Dapsone can be used as a steroid sparing agent with an

initial dose of 1mg/kg/ day.[3] Other drugs include mycophenolate mofetil and azathioprine.[1]

Rituximab is an effective adjuvant therapy for the management of resistant cases.[3,14,15]

Lins Filho et al reported childhood pemphigus vulgaris in a 8 year old boy with oral, conjunctival and genital ulcers. He was treated with prednisolone and dapsone as in our case.[3] Surya V et al reported 2 cases of childhood pemphigus with painful oral erosions without any cutaneous lesions. Both the patients were treated with systemic steroids in a tapering dose and topical triamcinolone.[1]

Though complete remission is rare, the recurrences can be reduced with early diagnosis as well as early initiation of treatment.

Conclusion

Childhood pemphigus vulgaris should be kept as a differential in all atypical vesiculobullous disorders and in children with toxic epidermal necrolysis like presentation. Same patient may present with various clinical presentations over a period of time. The disease tends to have a chronic course with multiple recurrences and hence long time follow up of each case is necessary.

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Table 1: Differential diagnosis for childhood pemphigus vulgaris.

Differential Diagnosis for childhood Pemphigus Vulgaris
Mucocutaneous diseases
Linear IgA disease
Epidermolysis Bullosa
Childhood Pemphigoid
Paraneoplastic pemphigus
Cicatricial pemphigoid
Predominant oral mucosal involvement
Recurrent aphthous stomatitis
Herpetic gingivostomatitis
Predominant genital mucosal involvement
Bullous fixed drug eruption
Sexual abuse

Figure 1,2 & 3

Figures 1 showing sheets of cutaneous erosions with superficial crusting and few areas of necrosis over face, neck, upper trunk with oral candidiasis. Figure 2 showing extensive erosions over posterior aspect of trunk, upper arms and gluteal region. Figure 3 showing erosions over the scrotum and healed hyperpigmented macules and plaques over lower legs.

Figure 4&5:

Figure 4 showing few discrete vesicles and crusted papules, few of them ruptured forming raw areas over the forehead. Figure 5 shows a healing erosion over the scrotum.

Figure 6:

Shows superficial erosions with epithelial debris over the anterior border of tongue, and an erosion near the angle of mouth towards right side.

Figure 7,8:

Figure 7: showing histopathological image ofsuprabasal blister with row of tomb stone appearance of basal keratinocytes. Figure 8shows intercellular deposition of IgG in the lower layers of epidermis.

Figure 9:

Shows multiple superficial erosions and crusting over the face, lips, neck and anterior aspect of trunk

Figure 10:

Shows multiple superficial erosions and crusting over posterior aspect of trunk

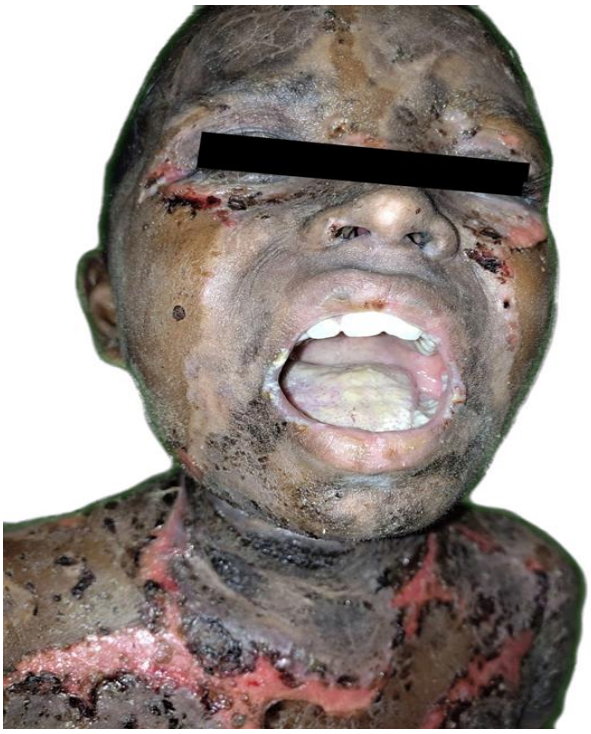

Figure 1: showing sheets of cutaneous erosions with superficial crusting and few areas of necrosis over face, neck, upper trunk with oral candidiasis.

Figure 2: showing extensive erosions over posterior aspect of trunk, upper arms and gluteal region.

Figure 3: showing erosions over the scrotum and healed hyperpigmented macules and plaques over lower legs.

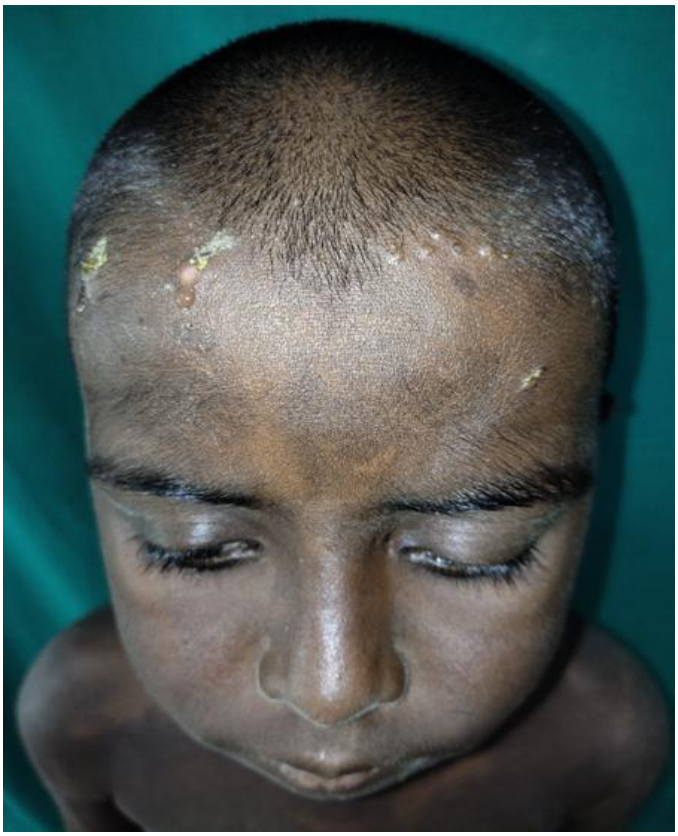

Figure 4: showing few discrete vesicles and crusted papules, few of them ruptured forming raw areas over the forehead.

Figure 5: shows a healing erosion over the scrotum.

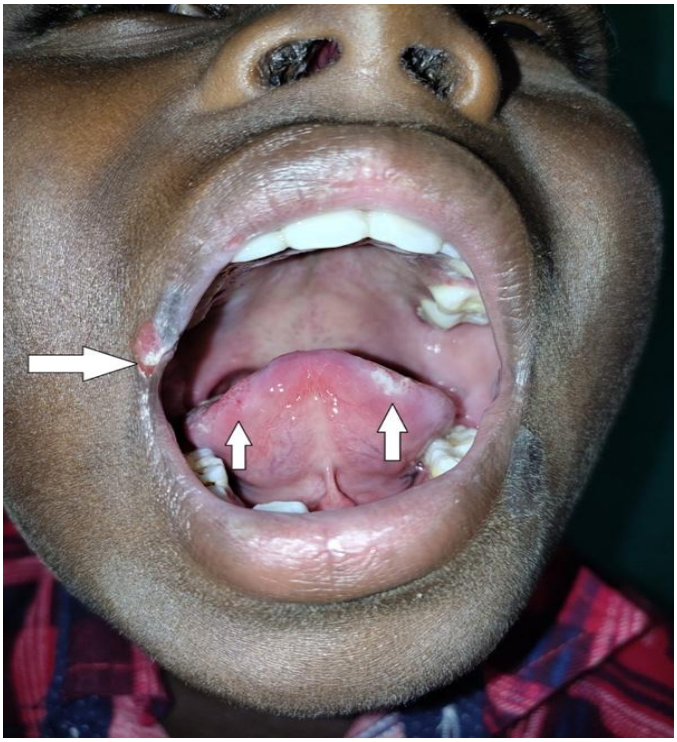

Figure 6: Shows superficial erosions with epithelial debris over the anterior border of tongue, and an erosion near the angle of mouth towards right side.

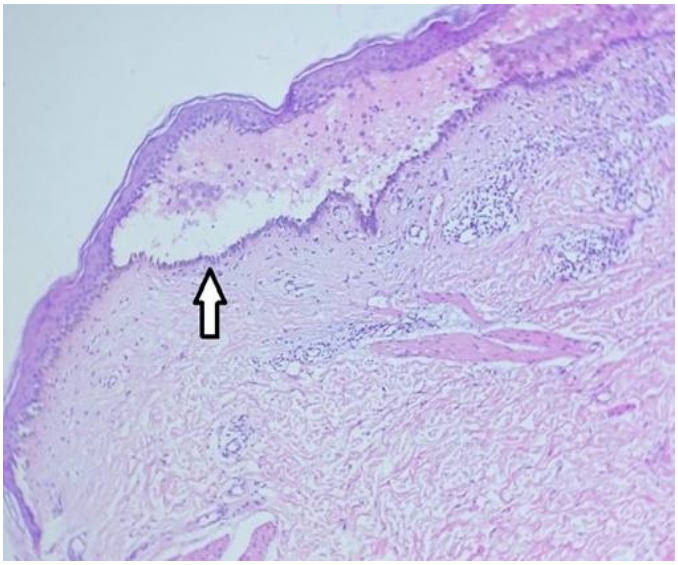

Figure 7: showing histopathological image of suprabasal blister with row of tomb stone appearance of basal keratinocytes.

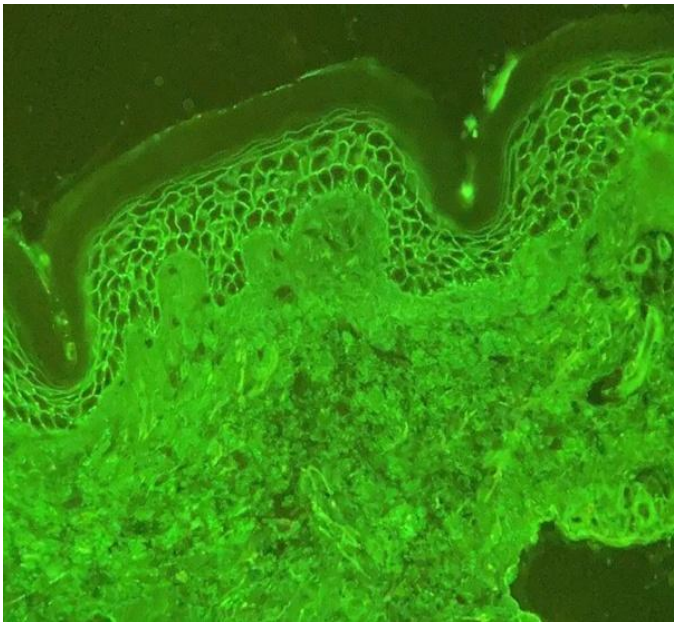

Figure 8: shows intercellular deposition of IgG in the lower layers of epidermis.

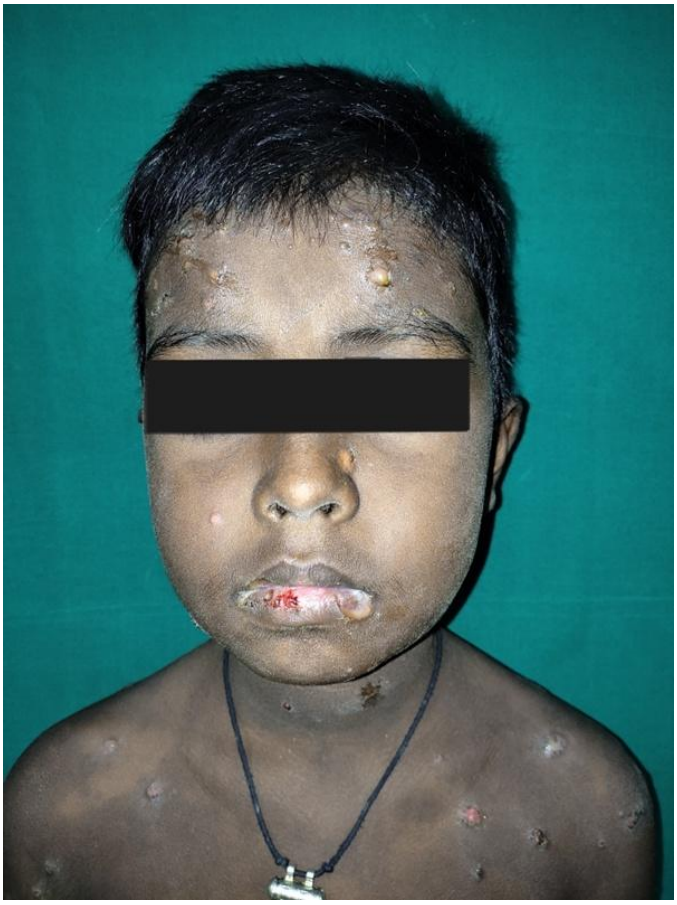

Figure 9: Shows multiple superficial erosions and crusting over the face, lips, neck and anterior aspect of trunk.

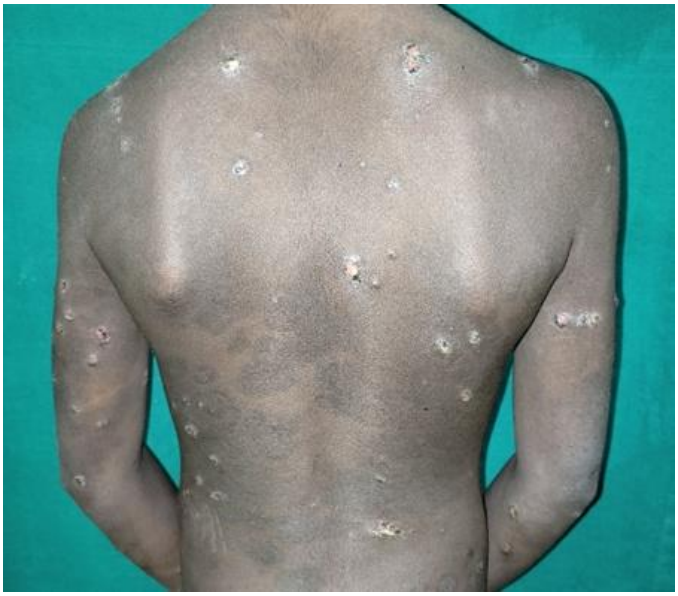

Figure 10: Show multiple superficial erosions and crusting over posterior aspect of trunk.