10.5005/jp-journals-10005-1434

CASE REPORT

Childhood Occurrence of Pemphigus

[1]Raju U Patil, [2]Rajesh T Anegundi, [3]Kumar R Gujjar, [4]KR Indushekar

ABSTRACT

Pemphigus is a chronic mucocutaneous disease that initially manifests in the form of intraoral blisters which spread to other mucous membrane and skin. This study describes an unusual case of chronic generalized childhood pemphigus disease in an 11-year-old girl, who presented with multiple vesicles all over her body. Such a condition is seen more often in older people rather than children. It is crucial for dental professionals to be familiar with the diagnosis of bullous skin diseases in children and adolescents, especially in its initial stages in order to prevent the serious consequences and morbidity. The article highlights clinical presentation, histopathology, and successful management strategies useful for pediatric dental practice.

Keywords: Acantholysis, Autoimmune, Blistering disease, Corticosteroids, Pemphigus.

How to cite this article: Patil RU, Anegundi RT, Gujjar KR, Indushekar KR. Childhood Occurrence of Pemphigus. Int J Clin Pediatr Dent 2017;10(2):196-200.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Blistering diseases are facing the danger of being finished, since our understanding of the pathogenesis and therapeutic approaches are undergoing a major revision. A wide spectrum of skin disorders can manifest as a blistering process. A blister is an event associated with tissue injury and fluid accumulation within a specific layer of skin due to either genetic mutations or autoimmune response. Blisters can also occur secondary to bacterial/viral infections, chemical/physical burns or skin necrosis/dermatitis.[1,2]

[1,2,4]Professor and Head, [3]Senior Lecturer

[1]Department of Pedodontics and Preventive Dentistry, Sinhgad Dental College & Hospital, Pune, Maharashtra, India

[2]Department of Pediatric Dentistry, SDM College of Dental Sciences & Hospital, Dharwad, Karnataka, India

[3]Department of Pediatric Dentistry, SEGi University, Kota Damansara, Selangor, Malaysia

[4]Department of Pedodontics and Preventive Dentistry, Sudha Rustagi College of Dental Sciences & Research, Faridabad Haryana, India

Corresponding Author: Raju U Patil, Professor and Head Department of Pedodontics and Preventive Dentistry, Sinhgad Dental College & Hospital, Pune, Maharashtra, India, Phone: +918871963896, e-mail: rupat13@yahoo.com

Here, the focus of interest is a bullous dermatoses in the child based on a case of pemphigus vulgaris (PV).

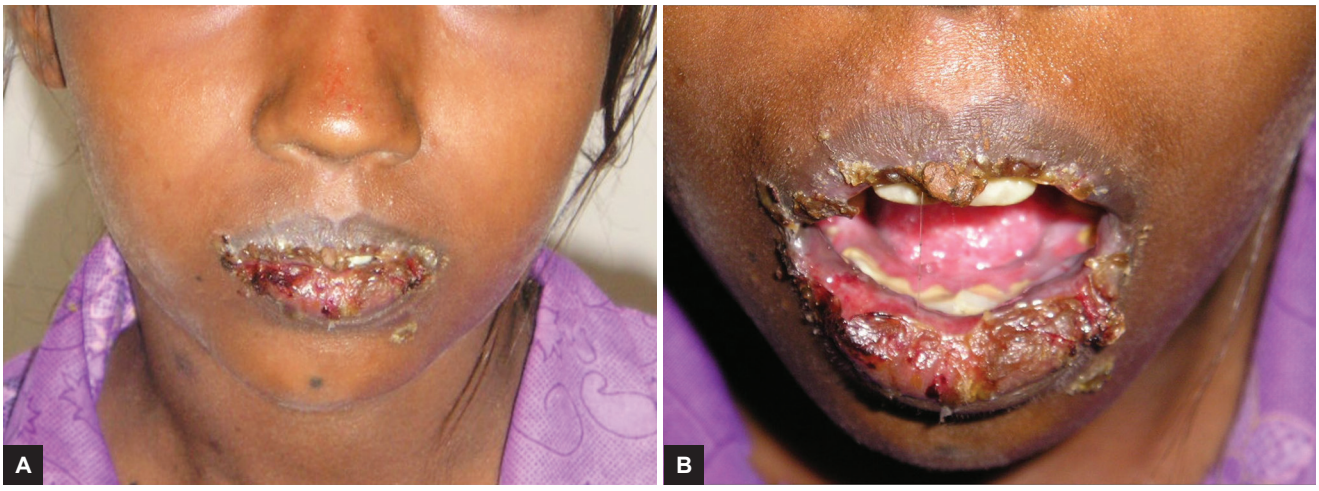
The PV is an autoimmune blistering disease of elderly (3rd-5th decade), which was previously fatal before the advent of steroid therapy, mainly due to dehydration or secondary systemic infection.[3,4] The PV is characterized by the presence of circulating autoantibodies immunoglobulin G against desmogleins 3[5,6] which result in loss of cell to cell adhesion and blister formation that rupture and progress to form painful erosions.[4] The PV in children aged less than 12 years is known as childhood PV and in those aged between 12 and 18 years as juvenile PV. Data on incidence and prevalence of childhood PV are scarce because in literature only a few cases are reported. In a study, children aged less than 15 years accounted for 3.7% of cases.[7] Several environmental factors, medications, and acantholytic substances superimposed on genetic predisposition may play a role in the onset of this disease in children.[8]

CASE REPORT

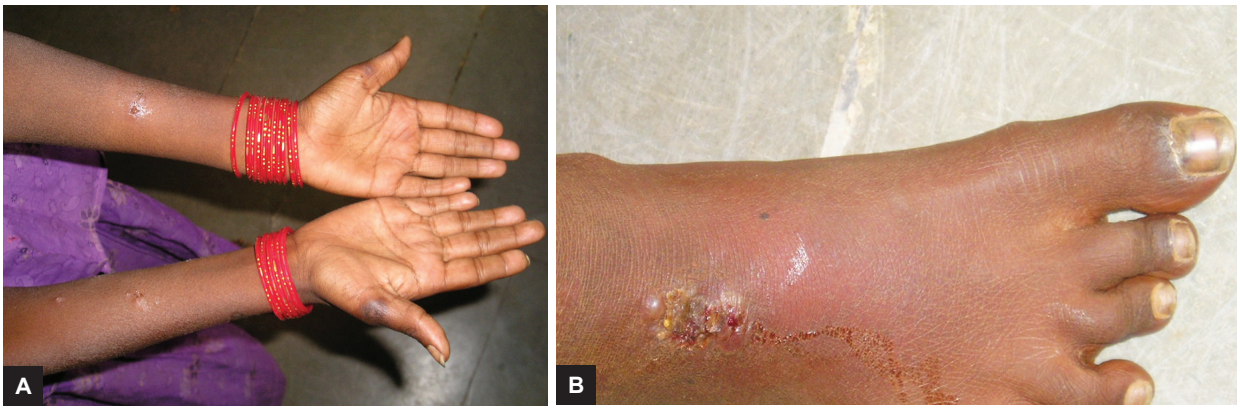
An 11-year-old girl presented to the department of pediatric dentistry, with a complaint of multiple eruptions and blisters all over the mouth, which increased in size gradually over a period of 2 to 3 months and ruptured to form a crusty erosive surfaces with watery discharge (Fig. 1). Later, similar sores appeared on limbs, trunk, and the genital area which were painful and led to considerable discomfort (Figs 2 and 3).

Entire oral mucosa including the tongue was eroded and erythematous, causing extreme discomfort and pain during eating. There was no history of any drug intake during the past 6 months nor any systemic condition identified. The child presented with such a condition for the first time and there was no such disorder noted in the family. Nikolsky's perilesional sign was positive.

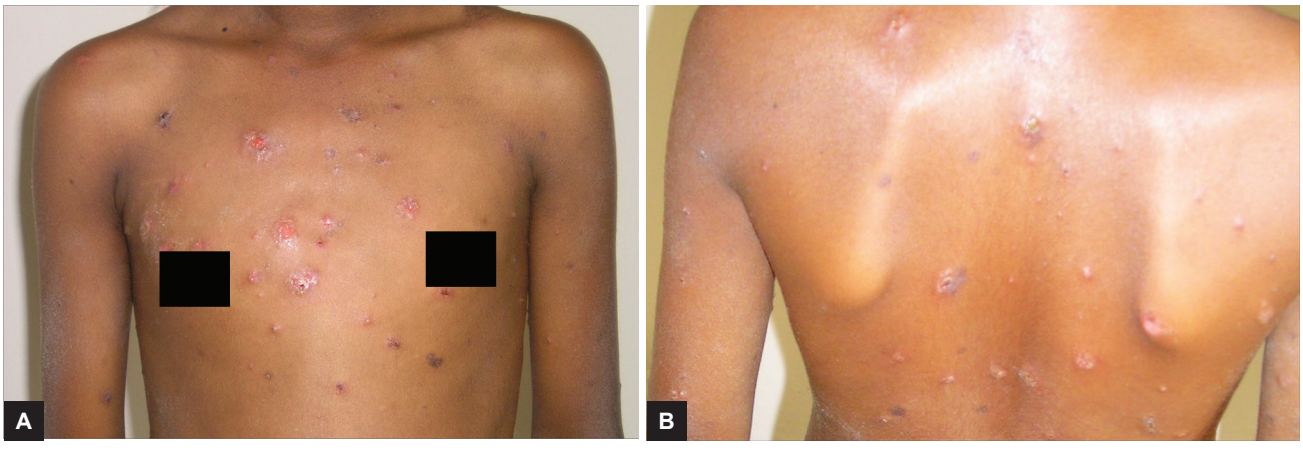
The girl was hospitalized in the medical unit and comprehensively managed with the help of a dermatologist (Tables 1 and 2). Direct immunofluorescence was positive and perilesion biopsy containing intact lesion, revealed Tzanck cells, intraepidermal blister and suprabasilar acantholysis (Fig. 4). The connective tissue stroma showed dense mononuclear infiltration. A significant improvement in the condition was observed after 3 to 4 weeks following the standardized steroid treatment regime (Figs 5 to 7).



Figs 1A and B: Multiple crushed lesions with superficial erosions on lips



Figs 2A and B: View of lesions on the limbs



Figs 3A and B: View of wide spread lesions all over the body

DISCUSSION

Unusual childhood occurrence, though quick response to treatment, however potentially life-threatening nature with substantial morbidity, justifies its consideration in routine dental practice. These chronic recurrent and painful lesions interfere with the daily activities of life, such as eating, drinking, talking, and personal relationships.[9] Pediatric dentists have the unique opportunity since initial lesions occur in the oral cavity and complete remission is possible only with early diagnosis.[10]

Prompt diagnosis and early initiation of aggressive therapy can combat the malignant course of disease in children. The treatment strategies should be based on the understanding of underlying pathogenic processes and

Table 1: Systemic treatment regime

Drugs	*Dose, route, and duration**	*Action*
Dexamethasone	0.5 mL Inj IM (50-100 mg) 3 to 4 weeks	Modification of immune response (immunosuppresion)
Roxithromycin	150 mg Tab BID – 2 to 3 week	Antibacterial for secondary infection
Prednisolone	10-20 mg Tab tapering to 5 mg BID – 2 to 3 months	Anti-inflammatory and modification of immune response
Hematopoietics	Oral capsule OD – 1 month	Nutritional supplement
NaCl saline	IV fluid	Electrolytic balance

*Minimum duration is 3 to 4 weeks, may be extended depending on response and recurrence; IM: Intramuscular injection; BID: Twice (two times) a day; OD: Once daily; IV: Intravenous

Table 2: Topical treatment regime

Drugs	*Dose, route, and duration**	*Action*
Triamcinolone	Local application for more than 3 weeks	Potent anti-inflammatory and alters immune response
Silver sulfadiazine and chlorhexidine	Local application for more than 2 weeks	Broad spectrum antimicrobial
Gentamycin with propyl salicylic acid	Local application for more than 2 weeks	Prevents secondary infections
Saline compresses over erosive lesions	Local application for more than 2 weeks	For soothing effect and control of edema
Chlorhexidine	Oral gargle for more than 3 weeks	Oral antimicrobial

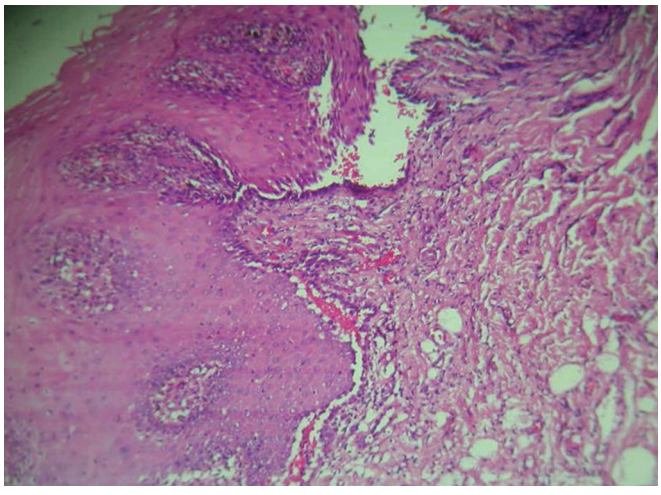


Fig. 4: Acantholysis and suprabasilar separation

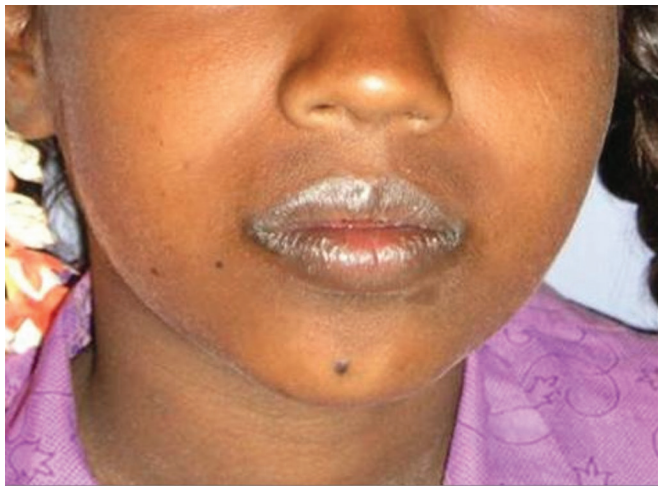
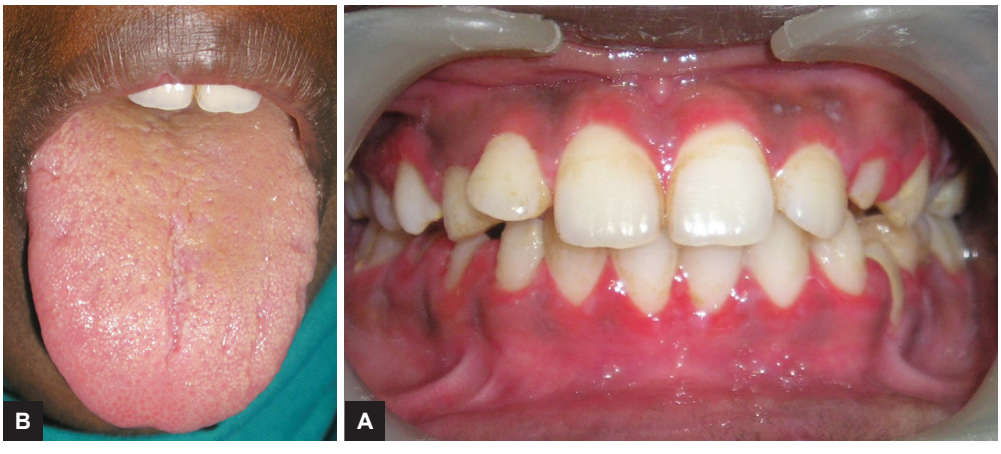
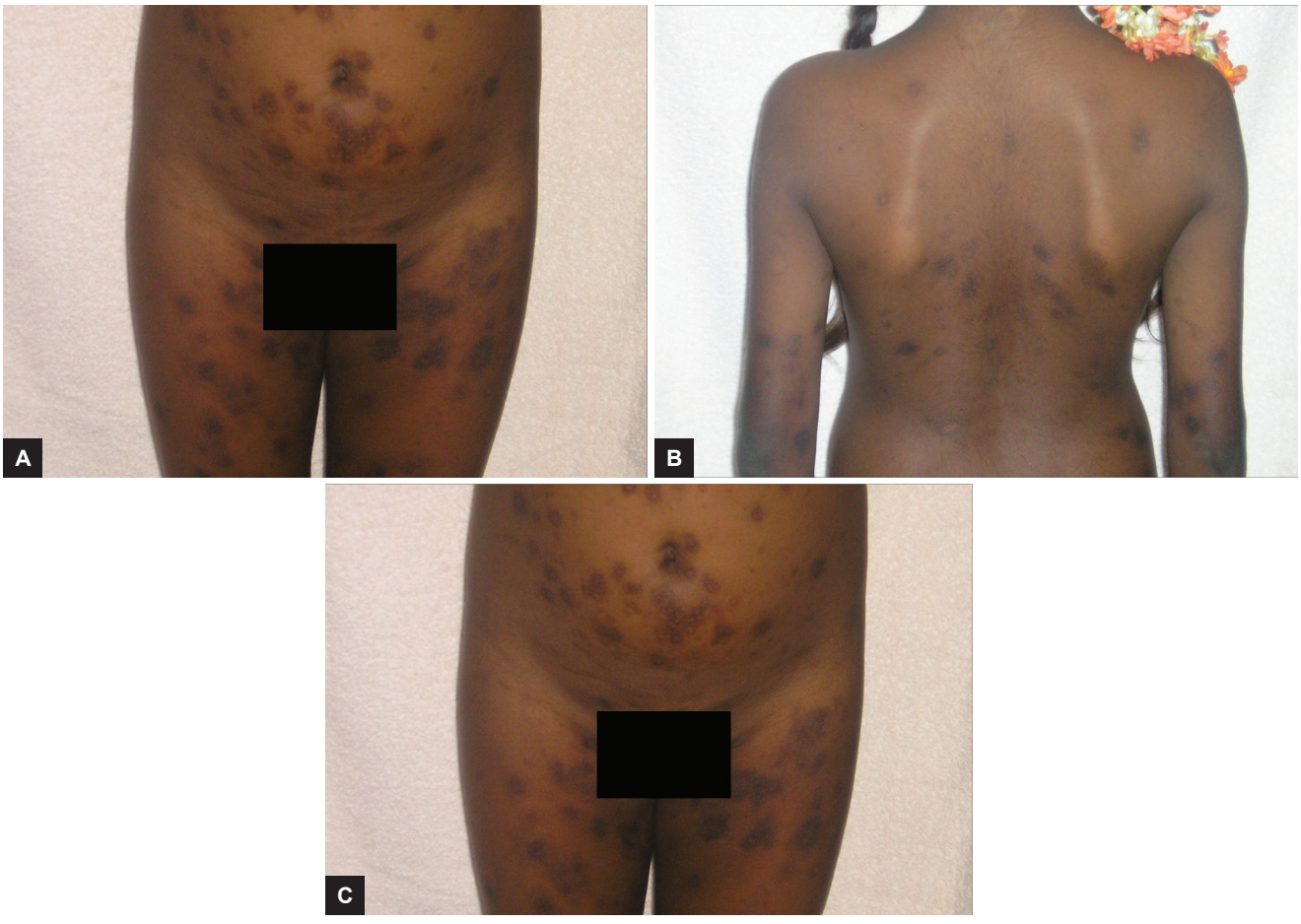


Fig. 5: Posttreatment view



Figs 6A and B: Lesions disappear following standard treatment regime

recurrence[3,11-14] (Tables 3 and 4). Systemic corticosteroids and immunosuppressive therapy are the mainstay treatments for PV. Apart from steroids, adjuvant therapies include azathioprine, mycophenolate mofetil, dapsone, and rituximab in refractory cases.[4,7,8] These modern therapies can effectively reduce the circulating antibodies, allowing patients to lead a normal life. Adverse effects associated with long-term use of steroids, such



Figs 7A to C: Healing of lesions all over the body

Table 3: Protocols for preventing recurrence[3,12-14]

Maintaining healthy diet and weight
Avoiding sunlight and friction of body folds
Keeping flexural areas clean and dry
Wearing cool garments with absorbent pads
Regular evaluation of secondary infections
Systemic antibiotics, such as tetracycline and erythromycin
Topical use of antibacterial creams, such as benzyl peroxide
Long-term low-dose steroid maintenance therapy
Controlling side effects of long-term steroids

Table 4: The bullous management portfolio[5,11-14]

Gold line mainstay of therapy – Steroids (Systemic prednisone 1 mg/kg/day and topical triamcinolone)
Broad-spectrum antibiotics for control of secondary infections
Improving the general health and hygiene of the patient (Fluid replacement, electrolytic balance, and multiple vitamins/minerals)
Symptomatic relief of pain, discomfort, burning, and itching (Paracetamol, astringents, and aluminium acetate)
Steroid sparing immunosuppressant and adjuvants (Mycophenolate mofetil, tracolimus, azathioprine, dapsone, retenoids methotrexate, cyclophosphamide, gold, cyclosporine, and chlorambucil)
Newer vistas - Plasmapheresis, intra venous immunoglobulins, anti-B cell monoclonal antibodies, CO_2 laser vaporization, dermabrasion, proteinase inhibitors, chimeric molecules, cholinergic agonists, etc.

as weight gain, menstrual irregularities, growth retardation, osteoporosis, and hormonal disturbances in adolescence[4,5] have always led to the search for newer steroid sparing and novel avenues for eradication of blisters at the molecular level.[1,2] As we probe deeper into molecular aspects of the disease, our understanding of the pathogenesis begins to gain focus, offering new novel, and improved methods of therapy or even an opportunity to achieve a cure, which should mark the end of an era of blistering diseases.

REFERENCES

1. Kalish RS. Pemphigus valguris: the other half of the story. J Clin Invest 2000 Dec 15;106(12):1433-1435.
2. Diaz LA, Giudice GJ. End of the century overview of skin blisters. Arch Dermatol 2000 Jan;136(1):106-112.
3. Robinson JC, Lozada-Nur F, Frieden I. Oral pemphigus vulgaris – A review of the literature and a report on the management of 12 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1997 Oct;84(4):349-355.
4. Toossi P, Hejazi S, Shakoie S, Asadi-kani Z. Childhood pemphigus vulgaris: a case report. Iran J Dermatol 2013;16(3):109-111.
5. Groves RW. Pemphigus: a brief review. Clinical Medicine 2009 Aug;9(4)371-375.
6. Amagai M, Koch PJ, Nishikawa T, Stanley JR. Pemphigus vulgaris antigen (Desmoglein 3) is localized in the lower

epidermis, the site of blister formation in patients. J Invest Dermatol 1996 Feb;106(2):351-355.
7. Kanwar AJ, Sawatkar GU, Vinay K, Hashimoto T. Childhood pemphigus vulgaris successfully treated with rituximab. Indian J Dermatol Venereol Leprol 2012 Sep-Oct;78(5): 632-634.
8. Lara-Corrales I, Pope E. Autoimmune blistering diseases in children. Semin Cutan Med Surg 2010 Jun;29(2): 85-91.
9. González-Moles MA, Scully C. Vesiculo-erosive oral mucosal disease--management with topical corticosteroids: (1) fundamental principles and specific agents available. J Dent Res 2005 Apr;84(4):294-301.
10. Sirois D, Leigh JE, Sollecito TP. Oral pemphigus vulgaris preceding cutaneous lesion – recognition and diagnosis. J Am Dent Assoc 2000 Aug;131(8):1156-1160.
11. Galimberti RL, Kowalczuk AM, Bianchi O, Bonino MV,Garcia Garcia A. Chronic benign familial pemphigus. Int J Dermatol 1988 Sep;27(7):495-500.
12. Fine JD. Management of acquired bullous skin diseases. N Engl J Med 1995 Nov 30;333(22):1475-1484.
13. Scully C, Challacombe SJ. Pemphigus vulgaris: update on etiopathogenesis, oral manifestations and management. Crit Rev Oral Biol Med 2002;13(5):397-408.
14. Solanki A, Chokshi K, Patel S. Review article on treatment of pemphigus. Int J Curr Pharm Res 2012 Aug 25;4(4):10-14.