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CASE REPORT

Juvenile Pemphigus Vulgaris

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ABSTRACT

Pemphigus vulgaris (PV) is a chronic autoimmune blistering disease of the skin and mucous membranes that affects older individuals. The disease rarely affects children and adolescent patients. The first manifestation of PV appears in the oral mucosa in the majority of patients, followed by cutaneous lesions. The diagnosis is based on clinical findings and laboratory analyses, and it is usually treated by the combined administration of corticosteroids and immunosuppressants. Dental specialists must be proficient to recognize the clinical features of PV to ensure early diagnosis and treatment, so that it determines the favorable prognosis and course of the disease. This article reports a case of juvenile PV.

Keywords: Immunofluorescence, Juvenile, Nikolsky's sign, Pemphigus vulgaris.

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INTRODUCTION

Pemphigus includes a group of autoimmune, potentially life-threatening diseases that cause blisters and erosions of the skin and mucous membranes. Pemphigus vulgaris (PV) is the most commonly observed member of a group of chronic autoimmune mucocutaneous diseases characterized by the development of intraepithelial blisters. It is an unusual disease (0.1–0.5 cases/100,000 inhabitants/yr), with onset in the 5th or 6th decade of life. The PV is infrequent in children and adolescents, but some cases have been reported in children and young adults; therefore, it should be taken into account in the differential diagnosis of children and young adults.[1]

A 14-year-old male patient came to the Department of Oral Medicine and Radiology with a chief complaint of pain and ulcers in his mouth since 6 months. He was apparently normal 6 months earlier; then he observed blisters in his mouth, which was sudden in onset and soon after a day, it ruptured to form an ulcer and the patient experienced pain, which was gradual in onset, localized, sharp, and severe, which aggravated on consuming hot and spicy food and was relieved on medication. Patient had also noticed bleeding from the ulcers. Patient gave history of five to six episodes of ulcers in the last 6 months. Patient also noticed blisters on his foot and hand since 3 months, and he visited a local doctor where he was prescribed some topical medication after which the lesions healed temporarily; the lesions used to recur after some days. Patient's family, personal history, and medical and dental history were noncontributory. On general physical examination, patient was moderately built and nourished for his age, conscious and well oriented with normal gait. All vital signs were within normal limits. Erosions and scars were noted on the right hand and left leg region (Figs 1 and 2). On extraoral examination, scars were present on the forehead region (Fig. 3). On intraoral examination, multiple ulcers were noted on the right and left buccal mucosa, upper and lower labial mucosa, ventral surface, and lateral borders of the tongue (Fig. 4) with positive Nikolsky's sign. The ulcers were measuring about 2 × 1 cm, irregular in shape; the floor of the ulcer was erythematous, margins were sloped, and edges were nonindurated. On the hard palate, an erythematous area was seen in respect to 16 region 2 cm lateral to midline measuring about 1 × 1.5 cm and irregular in shape (Fig. 5). Marginal and interdental gingiva were reddish is color (Fig. 6). Considering the history and the clinical features,

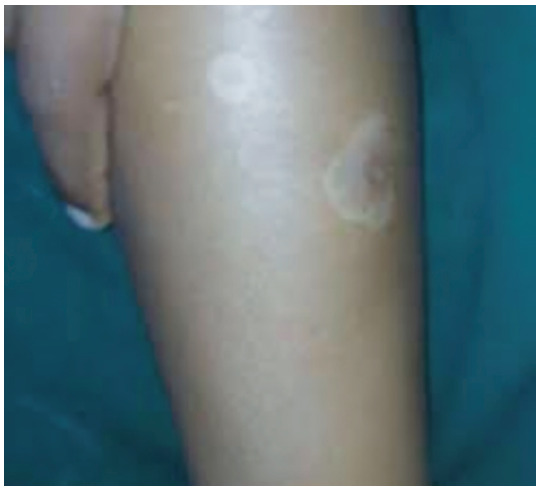

Fig. 1: Scars are seen over the posterior surface of left leg

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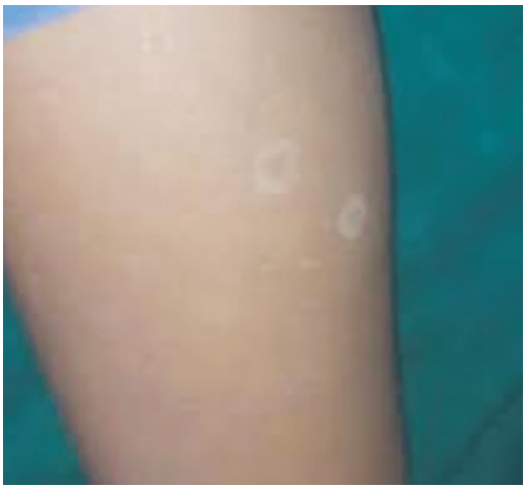

Fig. 2: Scars are seen over the posterior surface of right leg

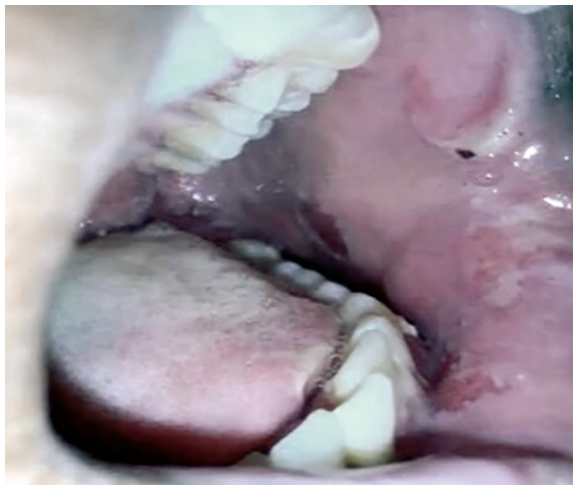

Fig. 4: Intraoral examination of left buccal mucosa

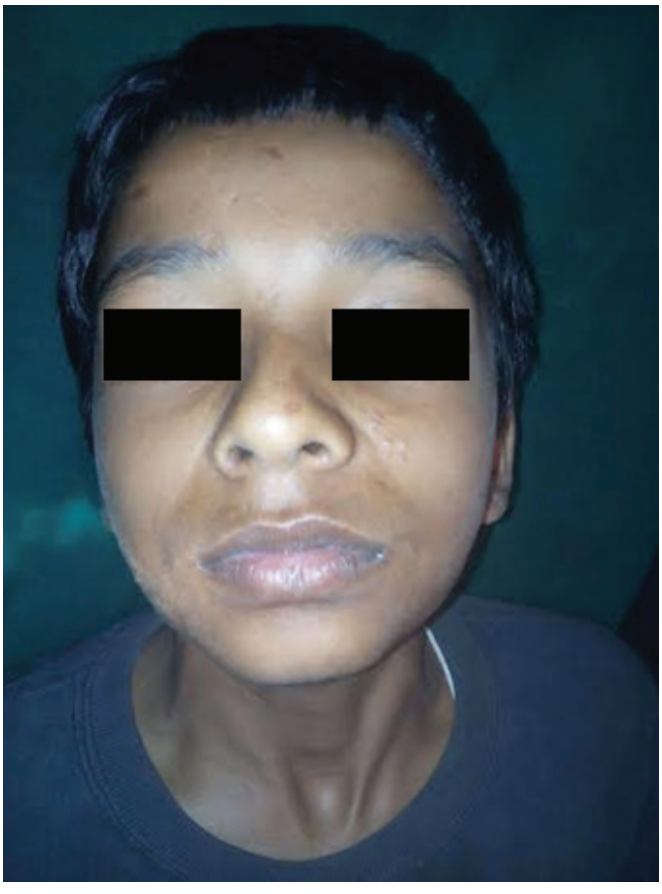

Fig. 3: Extraoral examination

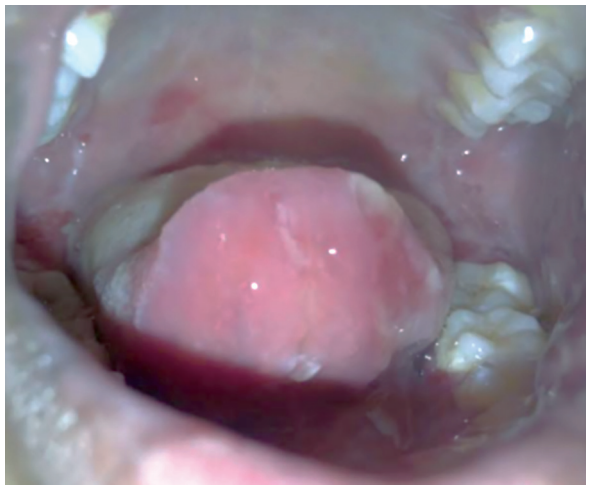

Fig. 5: Intraoral examination of hard palate

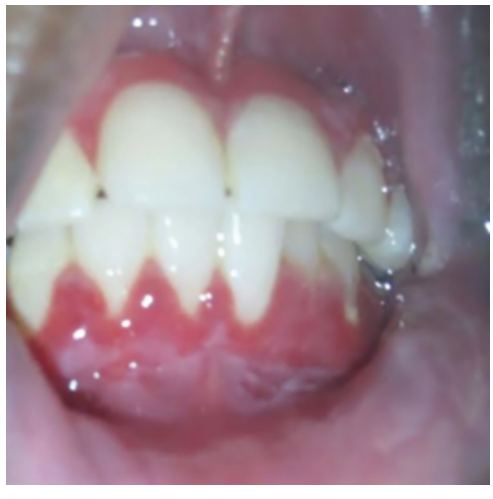

Fig. 6: Desquamative gingivitis

a provisional diagnosis of PV was given and differential diagnosis of bullous pemphigoid (BP) was considered. The patient was subjected to indirect immunofluorescence, which was done in 1:10 dilution in which esophageal antigen BP 120, BP 180, salt skin split test, and desmoglein 1 were negative and desmoglein 3 was positive. Photomicrograph showed immunoglobulin IgG accumulation in intracellular surface (Figure 7). These findings confirmed the diagnosis of PV. The patient was treated with topical steroids, namely triamcinolone acetonide 0.1% three to four times daily for a week along with topical analgesic like choline salicylate and topical anesthetic like lignocaine for oral lesions. There was slight reduction in the symptoms in the subsequent visit, and later after 10 days, patient again presented with severely progressed symptoms. Then patient was referred to a dermatologist

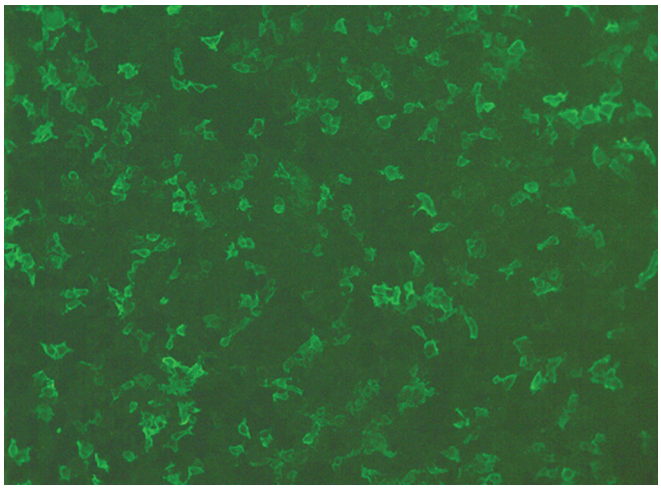

Fig. 7: Intercellular deposition of IgG antibody

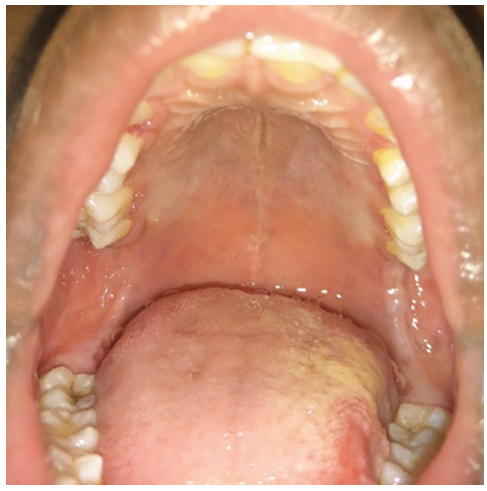

Fig. 8: Posttreatment picture of hard palate

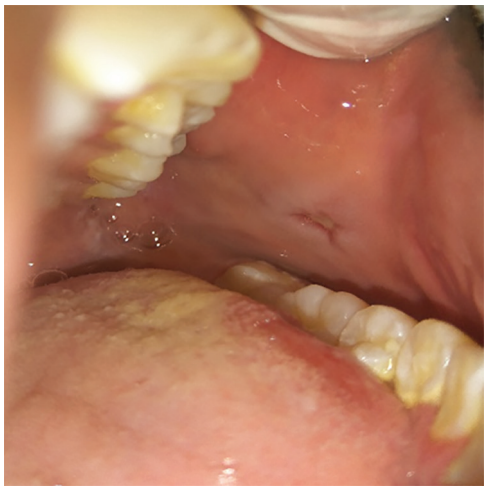

Fig. 9: Posttreatment picture of left buccal mucosa

where patient was put on immunosuppressive agent, IV rituximab 500 mg weekly for 1 month along with aloe vera gel. The oral lesions healed drastically (Figs 8 and 9) and the skin lesions healed with hyperpigmentation. The patient is on follow-up.

DISCUSSION

The term pemphigus is used to describe a group of chronic bullous disease originally named by Wichmann in 1791.[1] "Pemphix" in Greek means "bubbles or blisters" and "vulgaris" in Latin means "common." Pemphigus is a group of autoimmune blistering disorders characterized by blister formation, i.e., caused by loss of keratinocyte cell-to-cell adhesion in the epidermis due to circulating autoantibodies against desmosomal proteins.[2] The PV occurs mainly in 75% of the cases aged less than 45 years, and prevalence of pemphigus in childhood accounts for approximately 1.4 to 3.7% of total cases.

Pemphigus is a group of autoimmune blistering disorders characterized by blister formation, i.e., caused by loss of keratinocyte cell-to-cell adhesion in the epidermis due to circulating autoantibodies against desmosomal proteins. The initial lesions are often insidious and localized. There are 0.5 to 3.2 cases reported each year per 100,000 population, with the highest incidence in the 5th and 6th decade of life, with male-to-female ratio of 1:2. The mouth is affected by persistent, painful ulcers and a burning sensation, which affects the appetite. The skin becomes affected several weeks or months after the mucosal lesions appear, with the appearance of flaccid blisters filled with clear fluid. These fragile blisters are easily broken, which leaves behind erosions surrounded by epidermal rings. Putting pressure on healthy skin causes either a bulla or an erosion; this effect is known as Nikolsky's sign.[3,4]

Juvenile PV is a rare disorder, with a reported incidence of 0.1 to 0.5 cases per 100,000 individuals worldwide per year.[5]

It is slightly more common in girls and occurs primarily in adults during the 5th or 6th decade of life. The incidence of juvenile pemphigus is extremely low, so the diagnosis is often delayed due to rarity of pemphigus in this age group and confusion with other entities. So, high index of suspicion is required in order to make an early diagnosis. In children and adolescents, PV should be differentiated from erythema multiforme, acute herpetic gingivostomatitis, impetigo, linear IgA disease, epidermolysis bullosa, cicatricial pemphigoid, bullous pemphigus, and paraneoplastic pemphigus.[6] Histopathological and immunofluorescence examinations are very important in the diagnosis of pemphigus. Acantholysis and intraepidermal blister formation are characteristic findings on histopathological examination. Direct immunofluorescence shows the deposition of IgG around keratinocytes in a "chicken wire" or "crazy paving" pattern. The detection of circulating antiepidermal antibodies in the serum of patients with PV by indirect immunofluorescence further supports the diagnosis. The antibody

titers correlate with activity of disease in some patients and can be used for follow-up.[6] Since pemphigus is rare in pediatric population, evidence-based treatment guidelines have not been reported yet. Immunosuppression is the mainstay of therapy, and systemic corticosteroids are the treatment of choice (prednisolone 1–2 mg/kg/day) to control the disease during the acute phase.[6] In pediatric age group, the dose should be adjusted according to age, body weight, the severity of the condition, and the side effects of the drug. When the disease begins to go into remission, the dose can be tapered slowly. The continuation of treatment for a long time is not preferred due to long-term side effects of systemic steroids.[7]

The duration of therapies is shorter in juvenile PV patients than in adults for controlling disease.[8] The mortality rate in children (2.9%) is lesser than in adults (10–15%) due to this reduced immunosuppression.[8,9] Based on the rationale that pemphigus is primarily an autoantibody-driven autoimmune disorder, therapies that deplete autoreactive B-cell clones have been investigated for the treatment of pemphigus.[10] Rituximab, a monoclonal antibody directed against the CD20 antigen on B lymphocytes, has demonstrated efficacy for PV and pemphigus foliaceus.[1] Steroid is the first-line medication of PV, and rituximab is promising in refractory cases or steroid-sparing effect.

CONCLUSION

The diagnosis of pemphigus in children can be delayed due to the rarity of pemphigus in this age group, so a high index of suspicion is necessary for accurate diagnosis. The prognosis of disease seems to be better in childhood when compared with adults. The treatment of pemphigus is similar in children and adults, but clinicians should remember that children during periods of growth and development are extremely vulnerable to the side effects of systemic corticosteroid treatment.

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