Childhood pemphigus vulgaris: a case report

Parviz Toossi, MD
Somayeh Hejazi, MD
Safoura Shakoei, MD
Zahra Asadi-Kani, MD

Skin Research Center, Department of Dermatology, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Corresponding Author:
Safoura Shakoei, MD
Skin Research Center, Shahid Beheshti University of Medical Sciences, Shohada-e Tajrish Hospital, Shahrdari St, 1989934148, Tehran, Iran
E-mail: dr.shakoei@gmail.com

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Pemphigus vulgaris (PV) is a potentially fatal autoimmune mucocutaneous blistering disease. Although PV occurs predominantly in adulthood, in the 3rd to 5th decades of life, there have rarely been reports of childhood cases which are often misdiagnosed. It presents as oral blisters that rupture rapidly and progress to painful erosions. Most patients develop cutaneous flaccid blisters that rupture easily and leave painful erosions, which are slow to heal. These erosions are prone to secondary bacterial infection. Without treatment, the disease is progressive and the mortality rate is about 100%. We report an 8-year-old girl who was referred to our clinic from a pediatric center with mucocutaneous recalcitrant blisters.

Keywords: autoimmune bullous disease, childhood, pemphigus vulgaris

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INTRODUCTION

Pemphigus vulgaris is an autoimmune disease that occurs predominantly in adulthood. It is characterized by the presence of circulating auto antibodies directed against desmoglein 3 (Dsg3) with or without concomitant damage to Dsg1, which are desmosomal proteins responsible for keratinocyte adhesion. Autoantibodies result in loss of cell-cell adhesion and blister formation [1,2]. It presents as oral and cutaneous flaccid blisters that rupture rapidly and progress to painful erosions [1].

CASE REPORT

An 8-year-old previously healthy girl of Persian descent was admitted to hospital in 2011 because of a 2-month history of oral painful erosions and hemorrhagic crust formation that led to odynophagia. Severe odynophagia resulted in a 3kg weight loss. The sores were noted to begin in the buccal mucosa with progressive involvement of the lips and gingiva. She had partial improvement after a course of antibiotics. Bilateral purulent conjunctivitis with a watery discharge and photophobia began soon after the oral involvement, and the patient was ultimately admitted to a pediatric hospital center. There was no history of fever, joint pain, abdominal pain, diarrhea or exposure to medications before the onset of the lesions.

On physical examination, she was ill. There were widespread erosions of the lips, gingiva, and buccal mucosa (Figure 1). Ophthalmologic examination revealed periorbital erythema and conjunctivitis with vesicles and pustules. No uveitis was detected on slit-lamp examination, and visual acuity was normal. She was treated with oral acyclovir and oral cloxacillin with the diagnosis of herpetic gingivostomatitis. For ocular lesions, she received ophthalmic drops of betamethasone, ciprofloxacin, and tetracycline. After 10 days, mucosal lesions deteriorated and skin target lesions measured 0.5 to 0.8 cm in diameter appeared on various parts

of the body (Figure 2). The patient was referred to our clinic. A biopsy and direct immunofluorescence specimens were taken from one of her arm lesions with the clinical differential diagnoses of pemphigus vulgaris and pemphigus paraneoplastica (Figure 3). Direct immunofluorescence was positive for Immunoglobulin (Ig) G antibodies and C_3 to intercellular substance (ICS) of epithelial tissues (Figure 4). Once the diagnosis of pemphigus vulgaris was established, the patient was treated with 40 mg of prednisone and azathioprine 50 mg daily. After 1 month, significant improvement was observed and therefore corticosteroid treatment was continued with a dose of 35 mg/day. The dose of prednisone was tapered 5 mg every month.

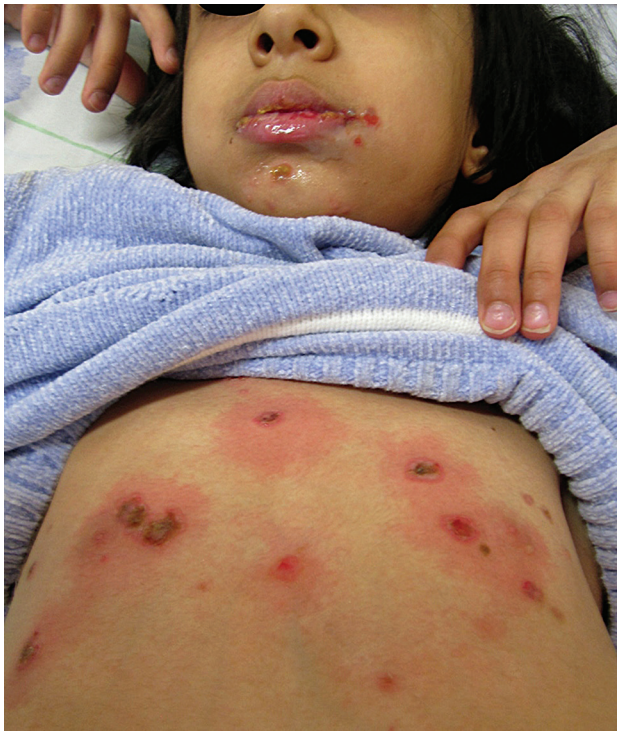

Figure 1. Abdominal flaccid blisters, and erosions and crusts of the lip

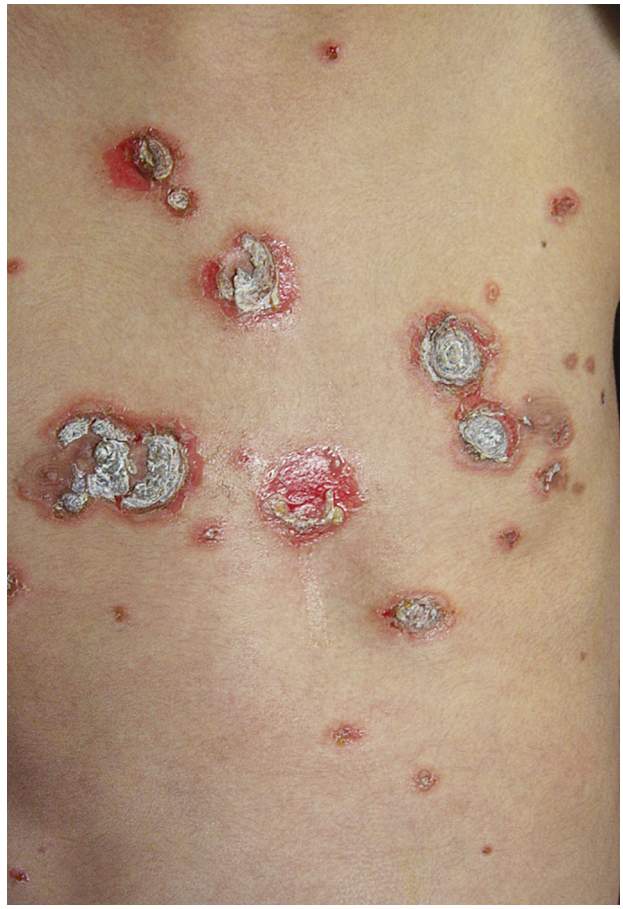

Figure 2. Flaccid blisters and target lesions of the trunk.

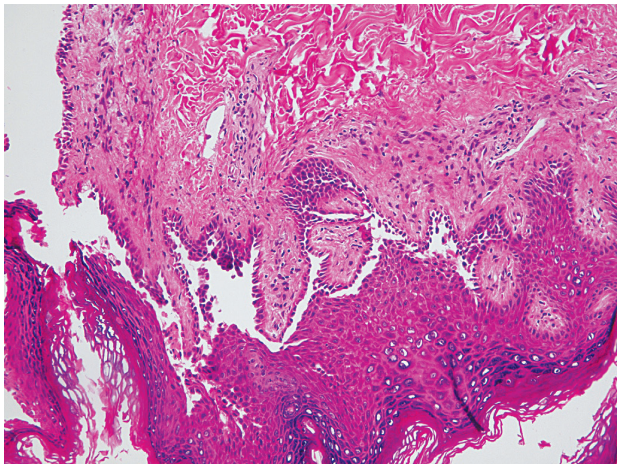

Figure 3. A Blisters in the skin show supra basilar acantholysis with a few acantholytic cells in the blister cavity (H&E ×40).

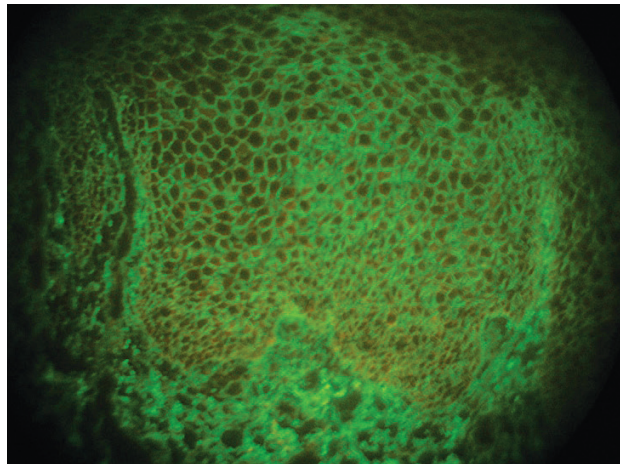

Figure 4. Direct immunofluorescence positive for IgG antibodies and C_3 to intercellular substance (ICS) of epithelial tissues.

DISCUSSION

Pemphigus vulgaris is an autoimmune mucocutaneous blistering disease [1]. The incidence rate of PV has been reported to vary between 0.42 and 1.62 cases per 100,000. The patients typically present in their 3rd to 5th decades of life [3-5]. The age at onset of juvenile pemphigus vulgaris (JPV) varies from 13 to 19 years (mean age: 14.9 years) [6]. The disease affects males and females equally in contrast to adult PV that has a slight female predominance [7]. In the majority of the patients, the skin eruption appears initially in the oral cavity. The lesions may be present on the gingiva, lips, buccal mucosa, and hard or soft palates. These lesions can progress to cutaneous involvement. The most common sites of cutaneous involvement in PV are the face, trunk, back, breasts, groin, and axillae, as described in our case [8]. The time period between the first symptoms and the diagnosis of JPV varies from 1 month to 2 years (mean: 8 months) [6]. Histological and immunopathologic features are similar to those seen in adults [6]. In one report, 30% of the patients had mild to moderate disease and 34% had moderately severe to severe disease [6]. Prior to the availability of corticosteroids, mortality in pemphigus vulgaris cases was very high.

The choices of treatment are corticosteroids. Adjuvant therapies such as azathioprine, mycophenolate mofetil, cyclophosphamide, and dapsone are added in case of a severe disease that cannot be controlled by corticosteroids alone or to reduce the dose of corticosteroids. Successful use of rituximab therapy has been reported in refractory childhood pemphigus vulgaris [9,10]. Use of intravenous immunoglobulin can delay the need for immunosuppressive drugs [9].

Adverse effects of therapy are reported in 19% of the patients which are mostly related to systemic corticosteroids. The reported incidence of adverse effects is as follows: infection, weight gain, cushingoid appearance, menstrual irregularity, hypertension, acne, cataract, growth retardation, osteopenia, diabetes mellitus, avascular necrosis, hormonal changes, physical and mental growth retardation, and social or cultural development arrest that occur during adolescence [6]. Weight gain and cushingoid appearance were seen in our case. Although systemic corticosteroids have been used to successfully treat the disease in most cases, long-term use is often necessary for adequate control. Newer therapies must be designed to adequately treat juvenile patients and limit serious adverse effects. The ratio of childhood cases was 2.9% in a large PV series. Therefore, we would like to emphasize that PV should not be neglected in the differential diagnosis of bullous eruption in children.

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