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CASE REPORT

Oral Ulcers as an Initial Presentation of Juvenile Pemphigus: A Case Report

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Pemphigus vulgaris (PV) is an autoimmune disease in which the autoantibody, immunoglobulin G, is directed against the keratinocytes in the epidermis. The classic presentations of PV are flaccid vesicles or bullae over the oral mucosa, trunk, groin, and extremities. The age of onset is usually between 40 and 60 years, and cases of PV in children or adolescent patients are rare. Here, we present a 17-year-old boy who had painful oral ulcers for 3 months initially and bullae spreading to the whole body in the following days. Paraneoplastic pemphigus was another differential diagnosis due to the atypical appearance of the skin lesion. However, PV was confirmed by hematoxylin and eosin staining and immunofluorescence examination of the skin biopsy specimens. The patient had a good response to corticosteroid treatment and the immunosuppressive agent, rituximab.

1. Introduction

Pemphigus vulgaris (PV) is an autoimmune blistering disease of the skin and mucous membranes. Autoantibodies directed against the surface of keratinocytes leads to the loss of cell-to-cell adhesion in the epidermis. The age of onset is usually between 40 and 60 years. The disease rarely affects children and adolescent patients.[1–4] PV is treated with high doses of systemic corticosteroids to control the painful and spreading lesions during the acute phase. Once the acute phase is controlled, immunosuppressive agents (ISAs) are used for the steroid-sparing effect and to reduce the production of pathogenic autoantibody. We herein report the case of a 17-year-old adolescent who had

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chronic oral ulcers initially and was diagnosed with PV based on the histopathologic and direct immunofluorescence (DIF) findings.

2. Case report

A 17-year-old, previously healthy, Taiwanese boy presented with a 3-month history of painful oral ulcers. Initially, he had been diagnosed with hand—foot—mouth disease. He received some supportive treatments such as analgesic drugs and local anesthetic spray but the effect was limited and the oral ulcers persisted. He denied any recent medication use or infections. There was no family history of autoimmune disease.

He had regular follow-up at the outpatient department of ears, nose, and throat. The oral ulcers persisted and refractory mucositis was impressed; therefore, prednisolone (0.35 mg/kg/day) was prescribed 1 month before this admission. However, its effect was limited. Furthermore, multiple vesicles arose from the lip, neck, hands, trunk, extremities, and groin to the buttocks. Because of these problems, he was referred to our outpatient clinic.

He had cough for 6—8 weeks, fatigue, and weight loss (4 kg), which were attributed to painful oral ulcers. He was underweight (body mass index: 16 kg/m^2) with pallid appearance. Facial examination revealed multiple oral ulcers with crust and necrotic tissue distributed from the outer lip to the soft and hard palate (Figure 1).

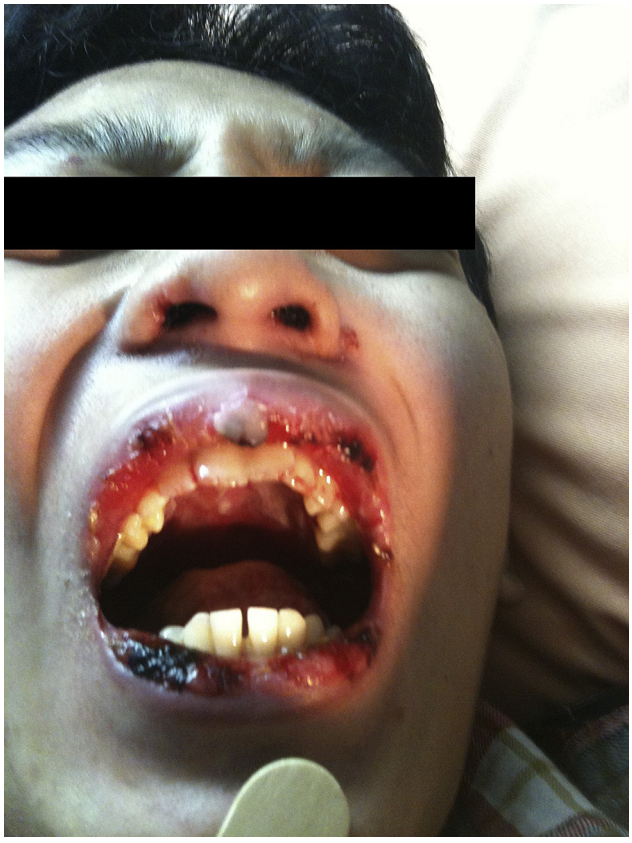

Figure 1 Oral ulcers with necrotic tissue on the top.

Furthermore, round vesicles and bullae with serous content, slightly tensed, were scattered across the trunk and four limbs (Figure 2). The initial differential diagnosis included herpes virus infection, human immunodeficiency virus (HIV) infection, Behçet's disease, and immune deficiency (Table 1). The Tzanck smear test was performed, which revealed a multinucleated giant cell.

Laboratory investigations revealed eosinophilia (9.2%) and lymphocyte-predominant white blood cell differentiation (50.4%), elevated erythrocyte sedimentation rate (18 mm/hour), elevated C-reactive protein (1.08 mg/dL), decreased immunoglobulin G (IgG) level (623 mg/dL), and negative titers for HIV screen, antinuclear antibodies, blood culture, herpes simplex virus-1 (HSV-1) IgM, HSV-2 IgM, and VZV IgM, but positive titer for varicella-zoster virus IgG. A chest X-ray showed increasing bilateral lung infiltration.

He received acyclovir (1500 mg/m^2/day) for the initial impression (chicken pox infection). Oxacillin (93 mg/kg/day) was also infused to cover the secondary bacterial infection. Intravenous Ig (IVIG; 976 mg/kg/day) was injected for 2 days for the low IgG level, which we considered might be related to the poor nutritional status over the past 3 months. However, the result was not so satisfactory.

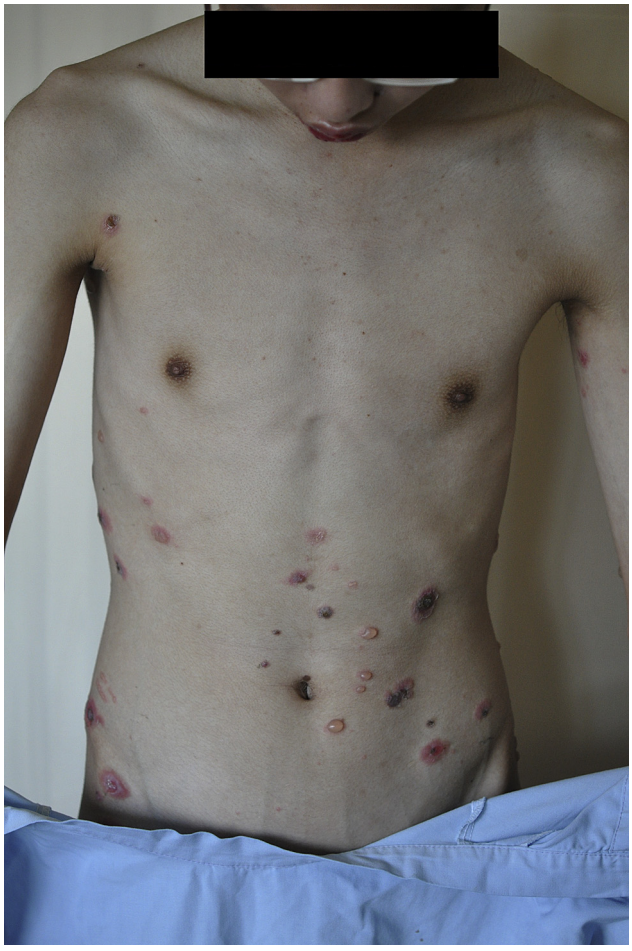

Figure 2 Vesicles and bullae spread to the trunk.