CASE LETTER

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Pemphigus Vulgaris in a Healthy Adult after COVID-19 Vaccination

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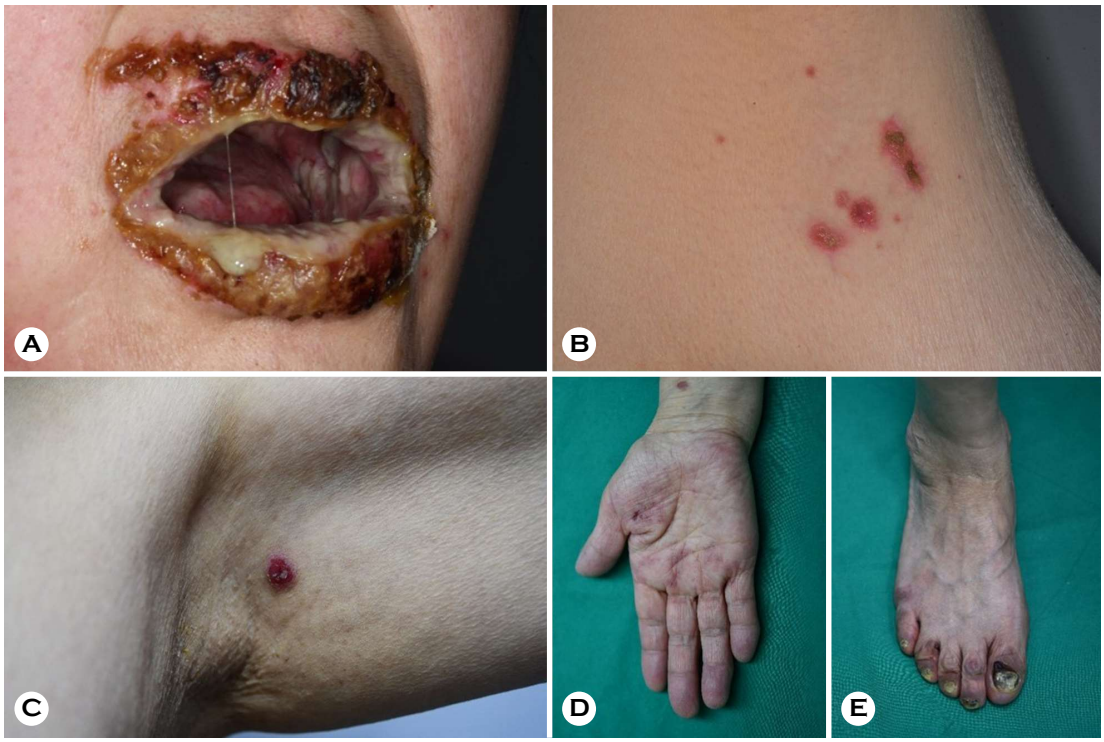

Fig. 1. Clinical presentations of the patient (A) Multiple severe painful various-sized erythematous eroded and crusted patches are observed on her cutaneous lips and the body of the lips with multiple oral mucosal ulcers. (B-E) Erythematous excoriated patches are found on the trunk, axilla, and acral areas. A flaccid blister is seen on the proximal nail fold of her right great toe.

Pemphigus is a group of skin autoimmune blistering diseases (AIBD) induced by anti-desmoglein 1 (Dsg1) and/or anti-desmoglein 3 (Dsg3). Although the precipitating factors, as with other autoimmune diseases, are unknown and are

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poorly comprehended, both genetic and environmental factors are known to impact its advancement. Regarding environmental factors, not only viruses but also vaccines such as influenza, rabies, and hepatitis B vaccines, can cause pemphigus as triggers. Recently, as the coronavirus disease 2019 (COVID-19) pandemic has spread globally, the number of COVID-19-vaccinated individuals is rising. Herein, we present a case of pemphigus following COVID-19 vaccination.

A 65-year-old woman presented to our clinic 3 days after receiving her second dose of the AstraZeneca® COVID-19 vaccine, with multiple painful, erythematous, eroded patches and pustules on the lips, oral cavity, trunk, axilla, and acral areas (Fig. 1). The lesion was also discovered on the anogenital area. Her first dose of the same vaccine was administered about a month ago. The patient had no serious medical history, including other autoimmune diseases. Laboratory

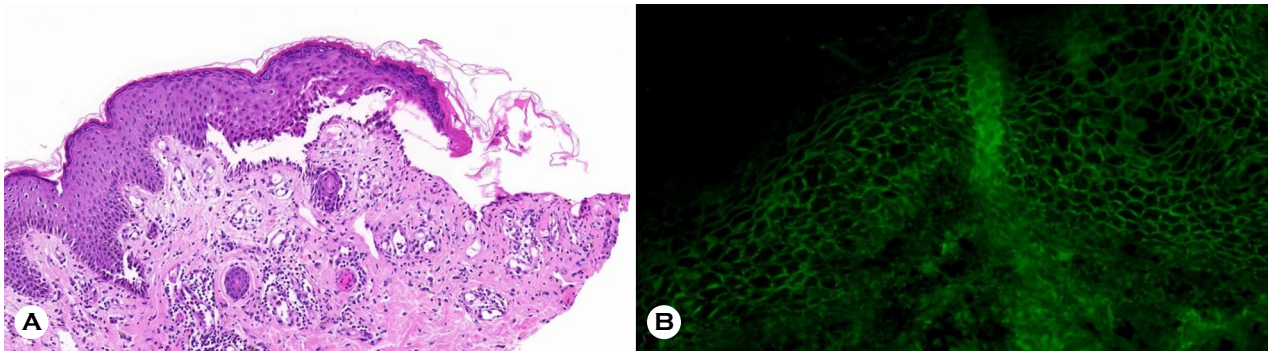

Fig. 2. (A) Histopathological examination of the skin demonstrates suprabasal acantholysis and a linear palisade of intact basal keratinocytes with a so-called 'row of tombstone appearance'. (B) Direct immunofluorescence (DIF) shows intercellular IgG deposition.

Table 1. Patients with pemphigus vulgaris after coronavirus disease 2019 (COVID-19) vaccination

No	Case report	Sex/ age	Location	Vaccine	Inoculation round	Histo pathology	DIF*	DSG1†/ DSG3‡
1	Thongprasom et al.[1]	F/38	Oral mucosa	Astra Zeneca	1st	Suggestive of pemphigus	Suggestive of pemphigus	N/A
2	Solimani et al.[2]	F/40	Oral mucosa, trunk and back	Pfizer	1st	Suprabasal acantholysis	IgG intercellular deposition	+/+
3	Koutlas et al.[3]	M/60	Oral mucosa	Moderna	2nd	Suprabasal acantholysis	IgG/C3 intercellular deposition	-/-
4	Knechtl et al.[4]	M/89	Oral mucosa, trunk, back, and left arm	Pfizer	2nd	Suprabasal acantholysis	IgG intercellular deposition	+/+
5	Calabria et al.[5]	F/60	Oral mucosa and oropharynx mucosa	Pfizer	2nd	Suprabasal acantholysis	IgG intercellular deposition	-/+
6	Our case	F/65	Oral mucosa, trunk, and anogenital and acral area	Astra Zeneca	2nd	Suprabasal acantholysis	IgG intercellular deposition	+/+

*DIF: Direct immunofluorescence; †DSG1: anti-desmoglein 1; ‡DSG3: anti-desmoglein 3

test outcomes were within normal limits except for a mildly elevated erythrocyte sedimentation rate (37 mm/h; normal (N) < 20 mm/h) and serum differential count of eosinophils (9.6%; N < 7%). The patient's serum was revealed to contain significant levels of anti-Dsg3 (398.5 U/mL; N < 7 U/mL) and anti-Dsg1 antibodies (154.2 U/mL; N < 14 U/mL) on an enzyme-linked immunosorbent assay. Histopathology revealed suprabasal acantholysis and a linear palisade of intact basal keratinocytes with a 'row of tombstone appearance' (Fig. 2A). Direct immunofluorescence (DIF) demonstrated intercellular IgG deposition (Fig. 2B). Indirect immunofluorescence found IgG deposition on the intercellular space at 1:40 dilution consistent with pemphigus Vulgaris. With a partial response, the patient was treated with prednisone 1 mg /kg and mycophenolic acid 30 mg/kg. She was moved to a different hospital for further evaluation and management.

Numerous studies have reported on pemphigus after COVID-19 vaccination (Table 1). All patients developed oral lesions several days after vaccination. Particularly, our patient also had lesions on the acral and anogenital areas. The patients were immunized against COVID-19 with AstraZeneca®, Moderna®, or Pfizer® COVID-19 vaccines. Except for one patient whose pathologic findings were not established, all histopathological and DIF examinations revealed suprabasal acantholysis and IgG intercellular deposition.

Virus- or vaccine-associated autoimmunity is a prevalent phenomenon, and multiple reports support a possible link between COVID-19 vaccination and new-onset AIBD like pemphigus, bullous pemphigoid, and linear IgA bullous dermatosis[5]. Except for one case, pemphigus after COVID-19 vaccination had a good prognosis overall[2], and complete remission can be achieved after weeks to months of standard treatment[1,3]. However, when receiving additional vaccinations, we should be watchful for recurrence or exacerbation of pemphigus[2]. With these potential side effects in mind, the association of COVID-19 vaccination with new-onset pemphigus must be confirmed. This case has the limitation of being a one-time occurrence, and the patient was lost to follow-up after transfer. Therefore, establishing a direct link was difficult. Additional research defining the relationship between autoimmune diseases and COVID-19 vaccination will be required to predict, prevent, and manage this vaccine's adverse effects.

Key Words: COVID-19 vaccination, Pemphigus vulgaris

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CONFLICT OF INTEREST

In relation to this article, we declare that there is no conflict of interest.

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PATIENT CONSENT STATEMENT

The patient provided written informed consent for the publication and use of her images.

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