Pemphigus Vegetans of Hallopeau: A Case Report

Dear Editor,

Pemphigus vegetans (PV) of Hallopeau is a rare and indolent variant of pemphigus clinically characterized by vegetating lesions preceded by pustules mainly in flexural areas (1,2). This helps us to differentiate it from PV of Neumann, which is a more extensive and refractory disease, more alike to a pemphigus vulgaris outbreak with blisters which turn into vegetating plaques (3). We report the clinical presentation, course, and therapeutic response in a patient diagnosed with PV of Hallopeau from its early stage during a 3-year follow up.

A 62-year-old man, non-smoker, presented at our clinic in July 2018 with hemorrhagic-serous crusts and fissures on the vermilion of the lower lip (Figure 1, a) and two merged circinate, sharply demarcated plaques on the right side of the groin (Figure 1, b). Plaque margins were elevated, with hypertrophic granulation tissue studded with pustules. Mucosal and cutaneous lesions persisted 6 and 4 weeks, respectively. The rest of the mucosa and skin were unaffected; the general state was good. The patient's family history for skin diseases was negative. The medical history included hypertension, atherosclerosis and hypercholesterolemia, hiatus hernia, and recent surgery (3 months prior) of an aortic abdominal aneurysm with reconstruction and synthetic graft placement. He was taking antihypertensives (fixed combination of 3 drugs, among them the ACE-inhibitor perindopril) with well-regulated blood pressure, statins, a pump-proton inhibitor, and acetylsalicylic acid. Differential blood count revealed eosinophilia.

Histopathology finding showed acanthosis, suprabasal clefting with a suprabasilar bulla and acantholysis, prominent eosinophilic intraepidermal spongiosis, and heavy dermal infiltration of eosinophils and lymphocytes (Figure 2, a and b). The diagnosis of pemphigus was confirmed by direct immunofluorescence (DIF), which detected C3 deposits on the surface of keratinocytes throughout the epidermis of perilesional skin. Circulating pemphigus antibodies were detected by indirect IF. Only Dsg 3 antibodies were detected using an ELISA assay (233.23 RU/mL).

After establishing the diagnosis of PV of Hallopeau, treatment with prednisolone 0.75 mg/kg/day orally in combination with adjuvant immunosuppression (azathioprine 100 mg daily) was started. Appropriate topical therapy with local steroids and antiseptic was applied. The steroid dose was titrated and gradually tapered down to the minimum required to control the disease – 10 mg. One-year remission was

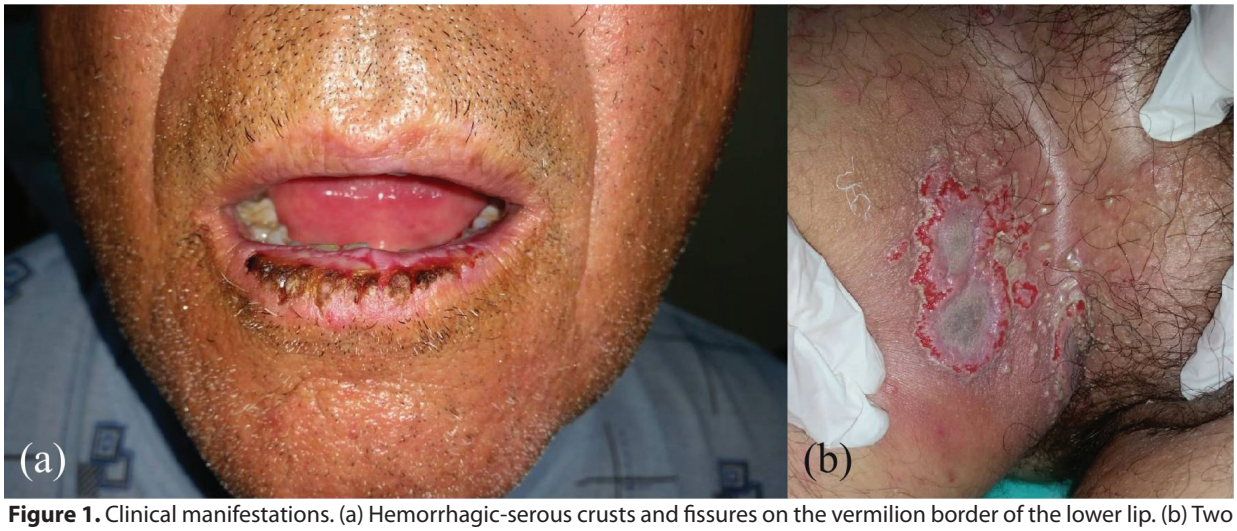

Figure 1. Clinical manifestations. (a) Hemorrhagic-serous crusts and fissures on the vermilion border of the lower lip. (b) Two merged circinate, sharply demarcated plaques with scattered pustules on the right side of the patient's groin.

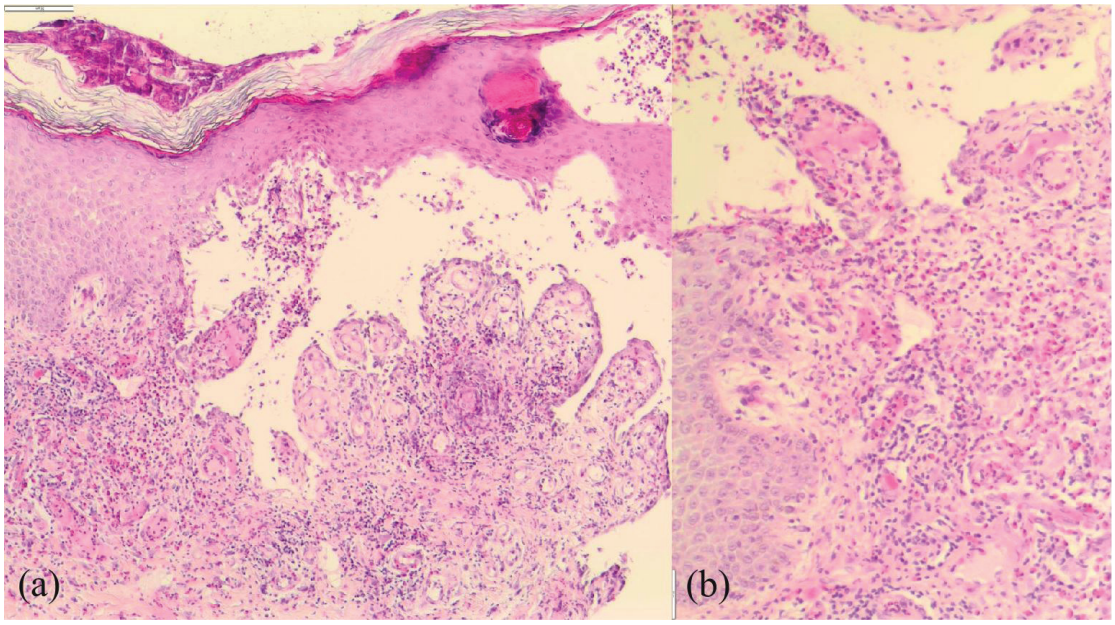

Figure 2. The histopathological findings (hematoxylin and eosin stain, ×20 and ×200, respectively). (a) Suprabasal clefting with a suprabasilar bulla and acantholysis, dermal infiltration of eosinophils and lymphocytes. (b) Prominent eosinophilic intraepidermal spongiosis and dermal infiltration with eosinophils.

achieved. Azathioprine was withdrawn in October 2019 and since then the patient experienced a flare-up twice. The control of pemphigus flare-ups was achieved by a low dose of steroids (30 mg prednisolone orally).

It remains debatable whether surgical trauma and radiology procedures such as angiographies (4) well as ACE-inhibitor drugs (5) triggered or aggravated the pemphigus.

Early recognition and correct diagnosis of this rare type of pemphigus allows us to treat and control the disease successfully with lower doses of steroids, reducing complications to the minimum.

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Jelena Maljić[1], Jaka Radoš[1], Davorin Lončarić[1], Ines Lakoš Jukić[1,2]

[1] Department of Dermatovenereology, University Hospital Centre Zagreb, Zagreb, Croatia

[2]University of Zagreb, School of Medicine, Zagreb, Croatia

Corresponding author:

Jelena Maljić, MD

University Hospital Centre Zagreb,

Zagreb, Croatia

jelena.kovac1991@gmail.com

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