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Case Report

Localized Bullous Pemphigoid in a Patient with Acquired Reactive Perforating Collagenosis

Masahiro Oka

Department of Dermatology, Kita-Harima Medical Center, Ono City, Japan

Keywords
Acquired reactive perforating collagenosis · Anti-BP180 antibody · Chronic renal failure · Diabetes mellitus · Bullous pemphigoid

Abstract
A 61-year-old man presented with 6-month and 5-day histories of multiple, pruritic nodular eruptions on the trunk and extremities and bullous eruptions on the left foot, respectively. The nodular eruptions had been treated with topical corticosteroids without improvement. He had been diagnosed with diabetes mellitus at the age of 42 years and had been suffering from end-stage renal disease for 1 year. Physical examination revealed scattered violet-brown papules and nodules on the trunk and extremities, many of which had central umbilicated necrosis or keratin plugs. Additionally, two tense bullae and five erosions were noted on the dorsal aspect of the left foot. Laboratory tests showed elevated levels of serum anti-bullous pemphigoid (BP)180 antibody. Histopathological findings of a nodule and a bulla were compatible with those of acquired reactive perforating collagenosis (ARPC) and BP, respectively. The papular and nodular lesions were diagnosed as ARPC, while bullous and erosive lesions were diagnosed as localized BP. The present case, together with previously reported cases of coexisting generalized BP and ARPC, suggests that coexistence of BP, regardless of whether generalized or localized, is significantly associated with ARPC.

Introduction

Bullous pemphigoid (BP) generally presents as widespread blister formation (generalized BP) [1]. Localized BP is a relatively rare variant of BP, in which the lesions develop only

Correspondence to:
Masahiro Oka, masahiro_oka@kitahari-mc.jp

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at localized sites [2]. Perforating dermatoses are a group of conditions characterized by transepidermal elimination (TEE) of dermal components, such as collagen, elastin, and fibrin [3]. Acquired reactive perforating collagenosis (ARPC) is a type of perforating dermatosis that usually develops in adulthood, in association with systemic diseases, especially diabetes mellitus or chronic renal failure [4, 5]. Recently, several cases of BP that developed in patients with ARPC have been reported [6–8]. However, no cases of localized BP developing in patients with ARPC have been described. Here, we describe such a case in a 61-year-old man for the first time.

Case Report

The patient presented with 6-month and 5-day histories of multiple, pruritic nodular eruptions on the trunk and extremities and bullous eruptions on the left foot, respectively. He had been diagnosed with diabetes mellitus at the age of 42 years and had been suffering from end-stage renal disease for 1 year. Physical examination revealed scattered violet-brown papules and nodules <2 cm in diameter on the trunk and extremities, many of which had central umbilicated necrosis or keratin plugs (Fig. 1a, b). In addition, two tense bullae and five erosions were noted on the dorsal aspect of the left foot (Fig. 1c). Laboratory tests showed elevated levels of serum anti-180-kDa BP antigen (BP180) antibodies (18.3 U/mL; normal, 0–8.9 U/mL). Serum levels of anti-BP230 antibodies were not measured. Histopathology of a biopsy specimen from a nodule revealed invagination of the epidermis with basophilic debris (Fig. 2a) and altered collagen fibers toward the external surface (Fig. 2b, c). Histopathological examination of a bulla demonstrated a sub-epidermal blister with moderate infiltration of eosinophils and lymphocytes (Fig. 2d). Direct immunofluorescence revealed linear deposition of immunoglobulin G and C3 along the basal membrane zone (Fig. 2e, f). The papular and nodular lesions were diagnosed as ARPC, while the bullous and erosive lesions were diagnosed as localized BP. After administration of oral prednisolone at 30 mg/day, the localized BP lesions rapidly resolved within a week. ARPC lesions also responded well to the therapy and almost resolved within 4 months. No recurrence of the bullous lesions was seen during the 8-month follow-up period.

Discussion

Various kinds of local stimulation, such as mechanical trauma and radiation, are known to trigger localized BP [2, 9]. Our patient had been wearing tight shoes and driving manual cars for his work since many years. Thus, his left foot had been subjected to repeated mechanical stress when he manipulated the clutch pedal with his left foot. Therefore, it is likely that the driving-induced mechanical stress on his foot was the stimulus for development of localized BP.

The major hypothesis regarding the mechanism of local stimulation-induced localized BP is that local stimulation might unmask BP antigens at the stimulated site, resulting in the production of antibodies against the BP antigen, leading to localized BP lesions [2, 9]. On the other hand, in cases with coexisting generalized BP and ARPC, some authors hypothesized that damage to the BMZ by scratching or TEE might generate BP autoantibodies against the BMZ through unmasking BP antigens, resulting in generalized BP lesions [6, 7]. Based on these hypotheses for the mechanism of localized BP or generalized BP in ARPC patients, we speculate that our patient already had low levels of anti-BP180 antibody by scratching- or TEE-induced unmasking of BP antigens when the patient suffered from ARPC, and local

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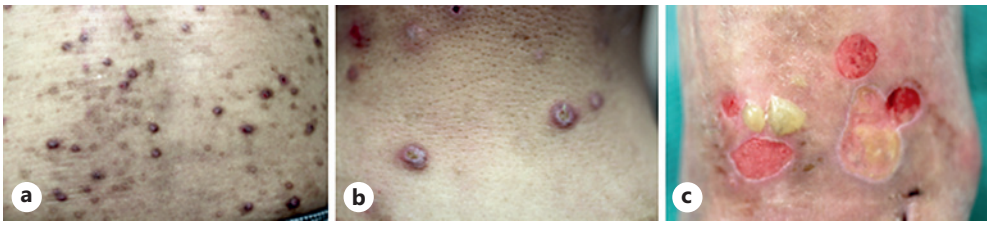

Fig. 1. Clinical appearance of the skin lesions. **a** Scattered violet-brown papules and nodules <2 cm in diameter were observed on the back. **a**, **b** Many of the papules and nodules had central umbilicated necrosis or keratin plugs. **c** Two tense bullae and five erosions were present on the dorsal aspect of the left foot.

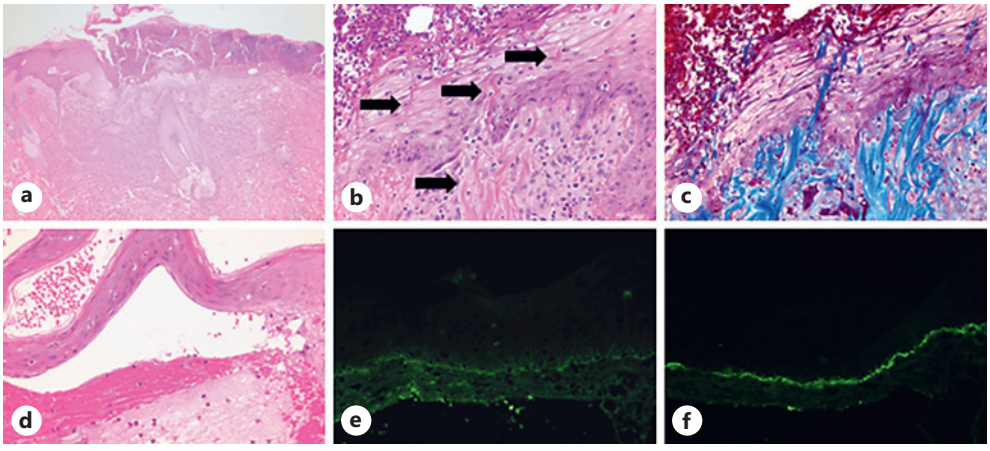

Fig. 2. a–f Histopathological findings of the skin lesions. A biopsy specimen from a nodule revealed a cup-shaped crust above the epidermis (H&E, original magnification, ×20; **a**) and altered collagen fibers toward the external surface (arrows) (H&E, original magnification, ×100; **b**). **c** The collagen fibers stained blue with Masson's trichrome (original magnification, ×100). **d** A biopsy specimen from a bulla showed a sub-epidermal blister with moderate infiltration of eosinophils and lymphocytes. Direct immunofluorescence showed linear deposition of IgG (**e**) and C3 (**f**) along the basal membrane zone.

stimulation on the foot while driving accelerated the unmasking of BP antigens, which led to increase in the levels of anti-BP180 antibody, resulting in localized BP lesions. We have recently reported a localized BP patient who was strongly suggested to have had anti-BP180 antibodies before the onset of localized BP [10].

In conclusion, the present case, together with the previously reported cases of coexistence of generalized BP and ARPC, suggests that BP, regardless of whether generalized or localized, is significantly associated with ARPC. The CARE checklist has been completed by the author for this case report, attached as online supplementary material (for all online suppl. material, see www.karger.com/doi/10.1159/000528140).

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and accompanying images. The study complied with the Declaration of Helsinki. This paper is exempt from ethical committee approval since we present a single case study.

Conflict of Interest Statement

The author declares no conflict of interest.

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The author did not receive any financial support for the present study.

Author Contributions

Masahiro Oka conducted the dermatological examinations of the patient, collected and analyzed data, treated the patient, and wrote the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author upon reasonable request.

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