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Case Report

A Case of Epidermolysis Bullosa Acquisita Triggered with Heavy Boots in Military Service •

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Abstract

Observations: Epidermolysis bullosa is a group of rare disorders which have in common the formation of blisters on minor physical injury, which are manifested in a variety of forms. It usually starts in childhood, but onset may be delayed until adult life.

A 20-year-old man with the complaint of blisters on his legs and arms since infantile period applied to our outpatient clinic. He expressed that his lesions exacerbated with heavy boots and shoes in military service. After taking a skin biopsy from the blisters histopathological, direct immunofluorescence and electron microscopic examinations were performed. Histopathologically, subepidermal blister formation, especially composed of lymphocytes and polymorphonuclear leukocytes, and inflammatory infiltration with perivascular edema in superficial dermis were observed. In addition to this, direct immunofluorescence examination revealed deposition of Ig G, Ig M, Ig A and C_3 on the floor of the blister. In electron microscopic examination, epidermis was found to be normal and there was epidermodermal separation. According to the clinical, histopathological, direct immunofluorescence and electron microscopic findings, the diagnosis was performed as epidermolysis bullosa acquisita.

Introduction

Epidermolysis bullosa is a group of rare disorders which have in common the formation of blisters on minor physical injury, which are manifested in a variety of forms [1, 2]. It is characterized by the development of vesicles and bullae over the joints of hands, elbows, knees, feet and other sites subject to repeated trauma [2].

Epidermolysis bullosa acquisita is a chronic, subepidermal blistering disease associated with autoimmunity to the collagen within anchoring fibril structures that are located at the dermal-epidermal junction [3, 4]. The features help to identify epidermolysis bullosa acquisita are skin fragility, predilection for traumatized areas [5].

Case Report

A 20-year-old man with the complaint of blisters and wounds on his legs and arms since infantile period applied to our outpatient clinic. He expressed that his lesions exacerbated with hot weather, heavy boots and shoes during military education in military service. His brother also had the same clinical findings.

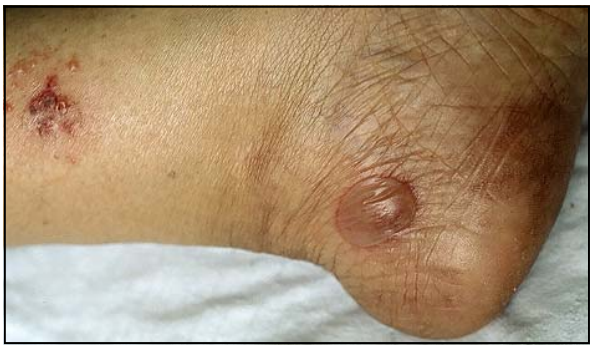

Figure 1. Vesicles and blisters localized to the left crural region

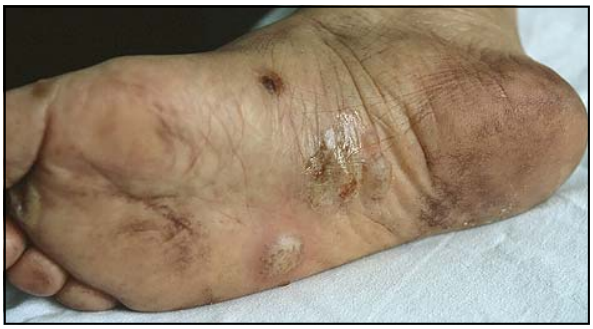

Figure 2. Blisters and exulceration localized to the right plantar region

Dermatological examination revealed vesicles, blisters and exulcerations localized to his foots and legs (**Figure 1, Figure 2**).

A skin biopsy was taken from a blister. Histopathological, direct immunofluorescence and electron microscopic examinations were performed. Histopathological examination revealed subepidermal blistering especially composed of lymphocytes and polymorphonuclear leucocytes, and inflammatory infiltration with perivascular edema on superficial dermis. In addition to this, direct immunofluorescence examination revealed deposition of IgG, IgM, IgA and C_3 at the floor of the blister. In electron microscopic examination epidermis was normal and there was epidermodermal separation (**Figure 3**).

According to the clinical, histopathological, direct immunofluorescence and electron microscopic findings, the diagnosis was performed as epidermolysis bullosa acquisita.

We applied sol de rivanol 0.1%, dexpanthenol and fucidic acid creams topically to the lesions. We also advised him to avoid major and minor traumas and prolonged sun exposure. In follow ups, clinical regression was observed.

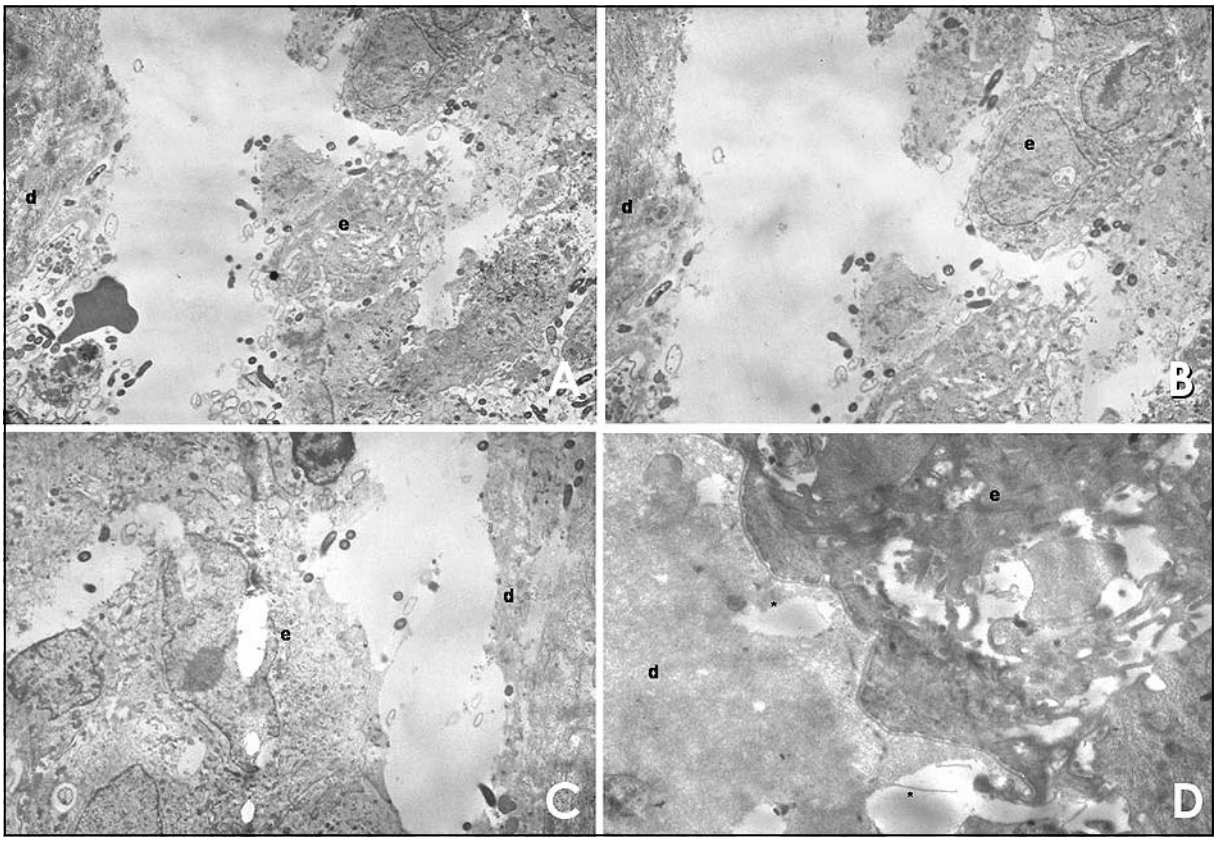

Figure 3A, B, C, D. Electron microscopic examination, blister formation between basal cells in epidermis and dermis, epidermodermal separation (**e**: epidermis, **d**: dermis) (*uranyl acetate peal citrate*).

Discussion

Epidermolysis bullosa acquisita is a very uncommon disease [5]. The classic form of epidermolysis bullosa acquisita is a mechanobullous disease marked by skin fragility. These patients have erosions, blisters over trauma-prone surfaces such as the back of the hands, knuckles, elbows, knees, sacral area and toes [3].

Routine histologic examination of lesional skin obtained from epidermolysis bullosa acquisita patients shows a subepidermal blister and clean separation between the epidermis and dermis. The degree of inflammatory infiltrate within the dermis usually reflects the degree of inflammation of the lesion observed by the clinician [3, 6].

Patients with epidermolysis bullosa acquisita have IgG deposits within the dermal-epidermal junction of their skin [7]. This is best detected by direct immunofluorescence of a biopsy specimen obtained from a perilesional site. IgG is predominant immunoglobulin class, but deposits of complement, IgA, IgM, factor B, and properdin may also be detected. The direct immunofluorescence staining demonstrates an intense, linear fluorescent band at the dermal-epidermal junction [3, 6].

With the advent of newer technology (*immunoelectron microscopy*, *Western immunoblotting* and *immunoprecipitation*), the diagnostic criteria for epidermolysis bullosa acquisita have become more precise. The common denominator for patients with epidermolysis bullosa acquisita is autoimmunity to type VII collagen [8]. The localization of the immune deposits within the dermal-epidermal junction of the skin of epidermolysis bullosa acquisita patients by immunoelectron microscopy is the gold standard for the diagnosis [3]. Histopathological, direct immunofluorescence and electron microscopic findings of our patient were associated with epidermolysis bullosa acquisita.

In general, the results of the treatment of epidermolysis bullosa acquisita are unsatisfactory. The noninflammatory types of bullous conditions are often best managed by supportive therapy [5].

Supportive therapy is warranted in all patients with epidermolysis bullosa acquisita. This includes instruction in open wound care and strategies for avoiding trauma. In some patients, it appears that prolonged sun exposure might aggravate or promote new lesions on the dorsal hands and knuckles. Avoidance of prolonged sun exposure and the use of sunscreens may be helpful. The patient should be educated to recognize localized skin infections and to seek medical care and antibiotic therapy promptly when this occurs [3]. Our case showed regression with supportive therapy.

Trauma-induced epidermolysis bullosa cases were described in the literature. *Tsianakas* et al reported a sporadic case of a patient suffering from bullous lesions induced by minor trauma on pretibial skin [9]. *Chao* et al described a 25-year-old male with easy blistering after trauma over the whole body from the age of 4 to 5 years. He was diagnosed as epidermolysis bullosa-*Köbner* type [10]. *Marr* et al reported of heat moulded footwear for a patient with epidermolysis bullosa [11]. Our case is interesting because his lesions were triggered with heavy boots and shoes in military service.

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