Case Report
Olgu Sunumu

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Giant variant of acquired perforating dermatosis in a patient with diabetes mellitus

Diabetes mellituslu bir olguda edinsel perforan dermatozun dev varyantı

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Abstract

Perforating dermatoses are a group of diseases characterized by transepidermal elimination of dermal components such as collagen, elastin and fibrin. Perforating dermatoses primarily include four main forms: reactive perforating collagenosis, elastosis perforans serpiginosa, perforating folliculitis and acquired perforating dermatosis. There is limited data about perforating dermatoses in the literature. Because of their rare appearance, they can be easily misdiagnosed. Acquired perforating dermatosis is a systemic disease such as diabetes and chronic renal failure; and it should be considered in case of pruritic papulonodules and giant plaques with central crater in a patient. In addition to the treatment of underlying disease in the first stage; narrow band ultraviolet B (nbUVB) is evaluated to be effective in these cases. Based on its rare occasions, we submit a giant variant of acquired perforating dermatosis with a collagen fiber predominant elimination pattern, which is treated with nbUVB.
Keywords: Perforating dermatosis, acquired perforating dermatosis, acquired reactive perforating collagenosis, phototherapy, giant variant

Öz

Perforan dermatozlar, kollajen, elastin, fibrin gibi dermal bileşenlerin transepidermal eliminasyonu ile karakterize bir grup hastalıktır. Perforan dermatozlar primer olarak reaktif perforan kollajenöz, elastozis perforans serpiginoza, perforan folikülit ve edinsel perforan dermatoz olmak üzere dört ana hastalık formunu kapsar. Perforan dermatozlarla ilgili literatürde az sayıda bilgi mevcuttur. Nadir görülmelerinden ötürü kolaylıkla dikkatten kaçabilmektedirler. Edinsel perforan dermatoz, özellikle diyabet, kronik böbrek yetmezliği gibi sistemik bir hastalığı olup kaşıntılı papülonodülleri, merkezi kraterli dev plakları olan hastalarda akla gelmelidir. Bu hastalarda öncelikli olarak alta yatan hastalığın tedavisine ek olarak dar bant Ultraviyole B'nin (dbUVB) etkili olabildiğini düşünmekteyiz. Biz de oldukça nadir görülmesi nedeniyle, dbUVB ile tedavi ettiğimiz, kollajen lif eliminasyon paterninin hakim olduğu dev lezyonlu bir edinsel perforan dermatoz olgusunu sunduk.
Anahtar Kelimeler: Perforan dermatoz, edinsel perforan dermatoz, akkiz reaktif perforan kollajenozis, fototerapi, dev varyant

Introduction

Perforating dermatoses are a group of diseases characterized by transepidermal elimination of dermal components such as collagen, elastin, and fibrin. Perforating dermatoses may occur as primary skin diseases characterized by transepidermal elimination (primary perforating dermatosis) or develop secondary to other dermatoses during their clinical course (secondary perforating dermatosis). Perforating dermatoses primarily include four main disease forms: reactive perforating collagenous (RPC), elastosis perforating serpiginosa (EPS), perforating folliculitis (PF) and acquired perforating dermatosis (APD). Of these, APD, usually associated with diabetes mellitus (DM) and chronic renal failure (CRF),

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includes Kyrle's disease, acquired RPC, acquired EPS and acquired PF. There is limited data about perforating dermatoses. We report a case of a giant variant of acquired perforating with collagen fiber predominant elimination pattern, which was treated with narrowband ultraviolet B (UVB), which has rarely been documented.

Case Report

A 63-year-old female patient with a 9-month history of pruritic crusted wounds on the trunk, arms and legs was admitted to our clinic. Her lesions had not improved with systemic antihistamine and topical corticosteroid treatments, which were given to her previously at other clinics. She was taking metformin and insulin for DM. On dermatological examination, she had a crusted ulcerated plaque with 6 cm in diameter on the outer quadrant of the right breast; a 2x3 cm crusted ulcerated plaque on the upper right quadrant of the abdomen, several hyperpigmented hyperkeratotic papules and scattered hyperpigmented macules on the anterior aspect of the trunk (Figure 1). The wound culture was negative. Histopathological examination revealed ulceration with fibrinous exudate (Figure 2), containing elastic fibrils (with Verhoeff staining) (Figure 3) and collagen bundles (with Masson's Trichrome staining) (Figure 4) in the epidermis. There were degenerated collagen bundles and mixed inflammatory infiltration in the superficial dermis and no abnormality found in deep dermis and subcutaneous tissue. Given these clinical and histopathological findings, she was diagnosed as giant variant of acquired perforating dermatosis with a collagen fiber predominant elimination pattern. She had normal hemogram, serum biochemistry, erythrocyte sedimentation rate, c-reactive protein, human immunodeficiency virus test, hepatitis markers, and chest X-ray. Narrowband UVB phototherapy three days in a week was started. After a total of 50 sessions, the lesions completely regressed (Figure 5). Informed consent was obtained from the patient for the use of photographs.

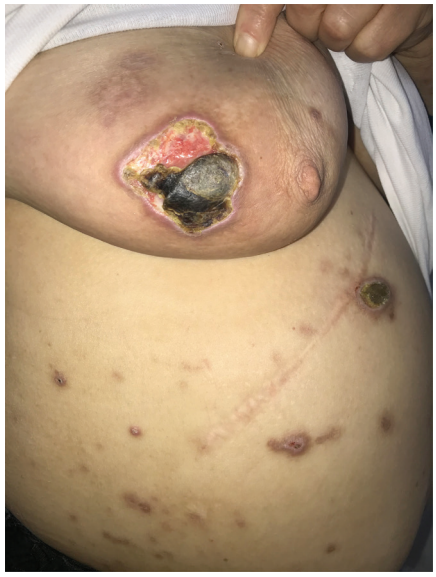

Figure 1. A crusted ulcerated plaque with 6 cm in diameter on the right breast; a 2x3 cm crusted ulcerated plaque on the abdomen, scattered hyperpigmented hyperkeratotic papules and hyperpigmented macules on the trunk

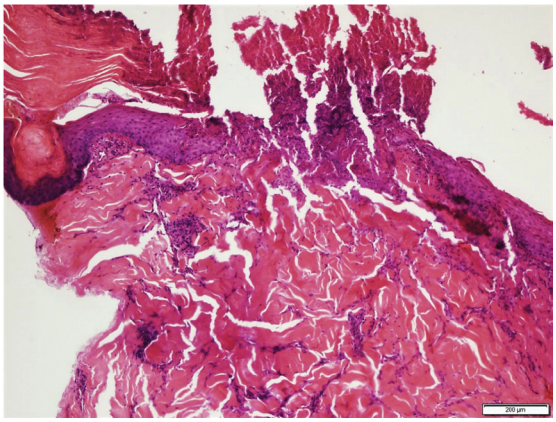

Figure 2. Epidermal ulceration, fibrinous exudate (original magnification x10, Hematoxylin & eosin)

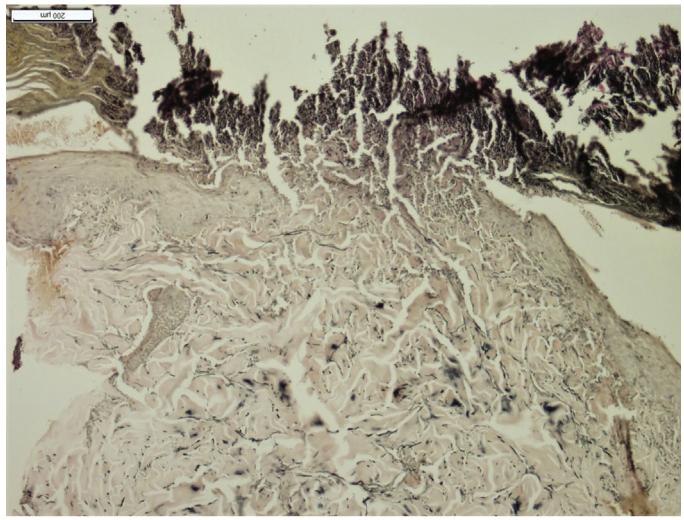

Figure 3. Epidermal ulceration, transepidermal elimination of elastin fibrils in the fibrinous exudate (original magnification x10, Verhoeff staining)

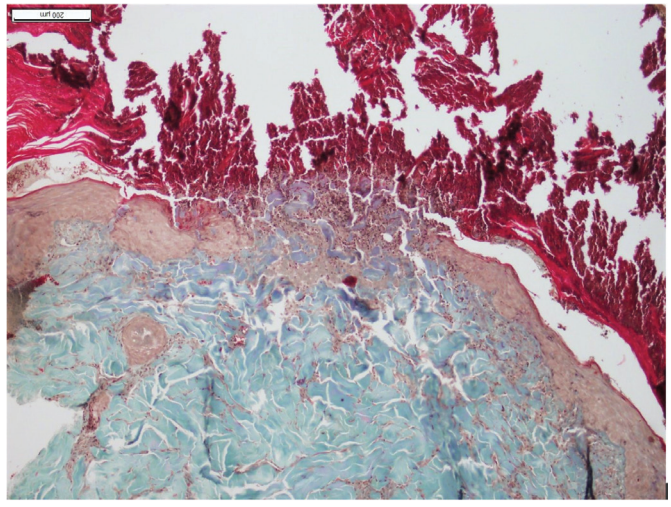

Figure 4. Epidermal ulceration, transepidermal elimination of collagen bundles (original magnification x10, Masson's Trichrome Staining)

Discussion

APD is an all-encompassing term that comprising various perforating diseases in adults, which is usually associated with an underlying systemic disease, particularly DM and CRF[1,2]. On the one hand, while the discussion of classification on this issue continues, on the other hand it is thought that APDs include acquired RPC, acquired EPS, acquired PF and Kyrle's disease. Acquired RPC is the most common form of APD, and Kyrle's disease, acquired PF and EPS are less common. APD may histologically resemble one of these four subtypes, but may also be manifested by overlapping of multiple elimination patterns. Since the manipulation of the lesions by the patients and the differences of the lesions may affect the pathological findings, the approach which includes all of these subtypes is more accepted than the approach which is classifying APD into histological subtypes[3].

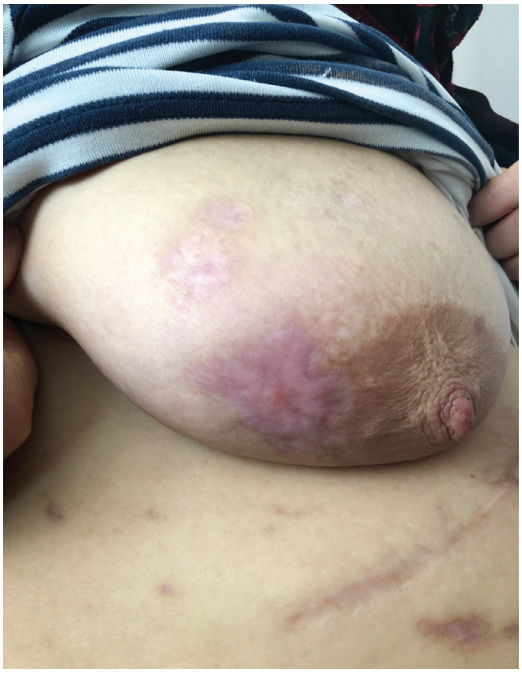

Figure 5. After treatment

The etiology and pathogenesis are unknown; thus it is controversial whether the issue is related to the epidermal or dermal area. Superficial trauma, diabetic microvasculopathy, calcium accumulation in the skin, and genetic predisposition are suggested hypotheses in the pathogenesis[3]. Since conveying substance from dermis through epidermis outside the skin is the main issue in the pathogenesis rather than active perforation of epidermis, the term "transepidermal elimination" is more accurate than "perforation"[4].

APD lesions usually present as umbilical hyperkeratotic papules with a size of 2-10 mm. The lesions are usually seen on the extensor surfaces of the lower extremities, but may also develop anywhere in the body, such as the trunk, scalp, conjunctiva and buccal mucosa[5]. Itching is the most common symptom, but pain is rare[6]. Koebner phenomenon may be observed, large plaques may be formed with the merging of papules on the itchy areas because of chronic trauma and itching. The largest lesion of our case was observed on trunk and had the size of 6 cm, which was larger than all other reported giant variant of APDs, including the first cases reported by Hoque et al.[7] (Table 1)[3,7-9].

APD usually arises in middle-aged adults associated with underlying diseases (Table 2)[10-22]. It is most commonly seen in patients with CRF on hemodialysis or DM[4]. In a study of 22 patients with APD; the most common associated diseases were CRF with 72.7%, DM with 50%,

Table 2. Systemic diseases associated with acquired perforating dermatoses

Diabetes mellitus[10]
Chronic renal failure[10]
Carcinoma (Liver[11], Hodgkin disease[12], colon[13], leukemia[14])
Endocrine diseases (Thyroid dysfunction[15], hyperparathyroidism[16])
AIDS[17]
Pulmonary aspergillosis[18]
Systemic lupus erythematosus[19]
Dermatomyositis[20]
Scabies[21]
Poland syndrome[22]

Table 1. Clinical features of reported cases of acquired perforating dermatosis giant variant

Case	Age, gender, race	Lesion localization	Lesion size	Associated disorders	Duration of diagnosis	Histopathological findings
1	77, F, Caucasian	Lower extremities	1-2 cm	Type1 DM	1 year	Like RPC
2	70, M, Caucasian	Trunk, extremities	1-2 cm	Type 2 DM, proteinuria, HT	3 months	Like RPC
3	37, M, Caucasian	Trunk, extremities	1-2 cm	Type 1 DM, ESRF, renal transplant	3 years	Like RPC
4	70, F, Caucasian	Trunk, extremities	1-2 cm	CRF, HT, breast cancer	5 months	Like RPC
5	57, M, Asian	Trunk, extremities, face	1-2 cm	Type 2 DM, CRF, hydronephrosis	5 years	Like RPC
6	60, F, Asian	Trunk, extremities	1-2 cm	ESRF in dialysis, hyperparathyroidism	6 months	Like EPS
7	60, F, Asian	Lower extremities	2,5 cm	Diabetic nephropathy	2 months	Like RPC
Our case	63, F, Caucasian	Trunk	6 cm	Type 2 DM	9 months	Like RPC

The first four cases (1-4) were reported by Hoque et al.[7], Case 5 by Gnanaraj et al.[9], Case 6 by Metterle et al.[3], Case 7 by Razmi et al.[10]
F: Female, M: Male, DM: Diabetes mellitus, CRF: Chronic renal failure, ESRF: End stage renal failure, EPS: Elastosis perforating serpiginosa, HT: Hypertension, RPC: Reactive perforating collagenosis

hepatitis with 27.3%, anti-hepatitis C antibody positivity with 13.6%, hypothyroidism with 9.1%, tuberculous lymphadenitis with 4.5%. In 13.6% of the cases, no association was found with any disease and all patients with CRF were receiving dialysis treatment[6].

The diagnosis of APD is based on clinical and histopathological evaluation. The histopathology of APD demonstrates a dermal lymphohistiocytic infiltration and a cup-shaped invagination with keratotic plug penetrating the papillary dermis. Orthokeratosis, parakeratosis or abnormal keratinization may be evident[7]. Transepidermal elimination of collagen, elastic fiber and degenerated follicular material can be seen as seen in typical RPC, EPS and PF, respectively. Basophilic debris is frequently seen in keratotic plugs[7].

Folliculitis, insect bite, prurigo nodularis, multiple keratoacanthoma, dermatofibroma, lichen plan, Darier's disease, verruca vulgaris, atypical mycobacterial infection should be considered in the differential diagnosis[3].

There is not any specific treatment for APDs, which have an optimistic prognosis. There may be some difficulties in treatment of APD as in there are some difficulties in distinguishing clinical subtypes. Treatments of APDs have not been evaluated in randomized controlled trials; treatments are mainly based on case reports or case series in the literature. First of all, treatment of the underlying disease may lead to improvement of APD. Besides that, antipruritic drugs, topical or intralesional corticosteroids, topical keratolytics, imiquimod, topical or systemic retinoic acids, methotrexate, allopurinol, phototherapy, and destructive methods such as excision, curettage, laser, cauterization, cryotherapy can be used as treatment for APD[3,7,23]. After a total of 50 sessions of narrowband UVB treatment, lesions of the patient completely improved.

Acquired perforating dermatoses may be easily overlooked because of their rare occurrence. Acquired perforating dermatoses should be considered especially in patients with a systemic disease such as diabetes, CRF or chronic skin disease such as atopic dermatitis with itchy papulonodules and giant plaques with central crater. The treatment of the underlying disease should be regulated, bathing with appropriate cleansers and keeping the skin moist, if any, itching should be reduced. In cases where these measures are inadequate, narrowband UVB treatment as in our case or the aforementioned treatment options can be used.

Ethics

Informed Consent: Informed consent was obtained from the patient for the use of photographs.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.Ö., Y.D., B.L., Concept: S.Ö., Design: S.Ö., A.K., Data Collection or Processing: S.Ö., Y.D., A.K., B.L., Analysis or Interpretation: S.Ö., A.K., Literature Search: S.Ö., Y.D., A.K., B.L., Writing: S.Ö., Y.D.

Conflict of Interest: No conflict of interest was declared by the authors.

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