Bullous systemic lupus erythematosus

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Bullous Dermatitis Artefacta

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Introduction and AIMS

Short Definition in Layman Terms

Bullous dermatitis artefacta is a mental abnormality in patients who mimic skin disease by inflicting themselves blisters. The diagnosis is immediately apparent to the doctor at first visit. The patient should be approached in such a way that the he is not losing face. Premature confrontation or embarrassing accusations should be avoided. Treatment strategy is narrow escape: the patient is almost confronted while he gets the chance to opt out by avoiding any scapegoat (coffee) or taking any rescue (vitamin C) that cleared the skin. In difficult cases the patient has to be confronted with the diagnosis by a psychiatrist.

Despite the spot diagnosis, DA needs a serious workup.

Learning Objectives

After reading this chapter you understand the clinical presentation, histopathology, differential diagnosis, and treatment approaches of bullous vesiculo-bullous eruptions in bullous dermatitis artefacta (DA).

Case Study: Part 1

A mother with child consulted the Center for Blistering Diseases after visiting three dermatologists before in the last year because of episodes of erosions in the face of the 12-year-old daughter. All dermatologists came to a prompt diagnosis of dermatitis artefacta, and ask the girl if she did it herself. She denied. The episodes persisted.

At dermatological examination I saw a shy but cooperative girl with linear erosions in the face with erythematous border. The mother was receptive for advice.

A skin biopsy for direct IF was negative.

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Didactical Questions; Cross Section of Questions to Prime the Readers Interest

What is the presentation of DA? What is the approach to avoid frustration of the doctor?

Facts and Figures

Definitions and Classification

Bullous dermatitis artefacta (DA) is a psychiatric factitious disorder in which the patient intentionally evokes blisters or erosions but denies self-infliction (Fig. 23.1). The synonym bullous pathomimia [1] is not used anymore, since not all patients are fully aware of their self-inflicting behavior that mimics bullous disease (Table 23.1). In automutilation (non-suicidal self-injury, DSM V) the patient also purposely wounds its own skin, but in contrast to DA admits self-infliction, such as cutting with a knife that does not mimic other skin disease. Neurotic excoriations are due to excessive compulsory scratching because of perceived itch.

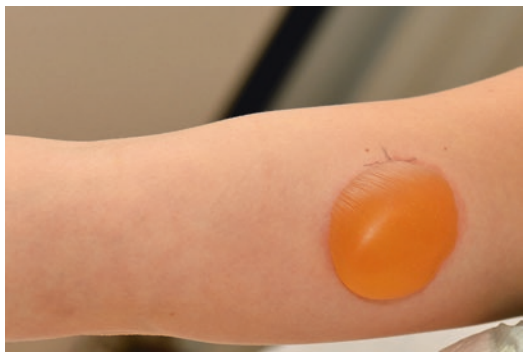

Fig. 23.1 Solitary monomorphic bulla on the arm of a teenager with bullous dermatitis artefacta. The level of blistering was subepidermal, and probably due thermally induced

Table 23.1 Diagnostic criteria for factitious disorders [3]

#	Criteria
1	Intensional production of physical or psychological signs or symptoms
2	Motivation for the behavior is to assume the sick role
3	Absence of external incentives for the behaviour (e.g., economic gain, avoiding legal responsibility, or improving physical well-being, as in malingering

Epidemiology

The patient with DA is predominantly female, and the bullous subtype mostly teenager. One of the parents, mostly the mother, is present at first consultation. The prognosis improves with younger patient, and shorter history of DA.

Pathogenesis

The patient keeps the secret of self-infliction or shares it with a relative (‘folie à deux’) or is the victim of a parent (Munchausen-by-proxy syndrome). The loneliness of the secret is compensated by the attention that the skin disease evokes in others. The patient may also not be fully conscious of the self-harm by dissociation. The psychopathology of this behavior is associated with border line personality disorder, multiple personality disorder, posttraumatic stress syndrome, anorexia and bulimia. Simply said: the patient dies for attention, but shows indifference for pain (‘la belle indifference’).

Diagnosis Paths

History and Physical Examination

Bullous DA is a spot diagnosis (Fig. 23.1): the physician immediately recognizes the bizarre pattern of the skin lesions, and considers artefacts. New lesions have developed “spontaneously” days before the first visit. The medical history is hollow with no timeline or evolution pattern. The patient appeals the competence of the doctor by questioning how these lesions suddenly can develop (Table 23.2).

General Diagnostics

Despite the spot diagnosis, DA needs a serious workup. This is important for a trustful patient-physician relation, but may also prevents to step into the pitfall of missing DA-like autoimmune blistering disease. I remember the case of a 58-year old female with a 10-year history of crusted erosions on arms and neck that healed with scars. She had visited three dermatologists, a rheumatologist and a psychologist who all presumed the diagnosis bullous DA. Taking a DIF biopsy turned out to be linear IgA bullous dermatosis. Treatment with dapsone cleared the lesions within weeks and saved her marriage.

The most important differential diagnosis of bullous DA is porphyria cutanea tarda and pseudoporphyria (Chap. 22). Therefore, take a biopsy, examine urine for uroporphyrines, and check history for culprit drugs. Limit the investigations and visits however, since that keeps the doctor to remain expert in the eyes of the patient.

The physician should take all efforts at the first visit to develop a trustful relation with the patient. Show genuine personal interest and ask the patient questions about social setting (home, school, sports). Address the accompanying person separately in the conversation. Do not let the patient loose face in anyway.

Case Study: Part 2

My spot diagnosis was dermatitis artefacta. The patient with the nurse were sent away for drinking thee, and I took the opportunity to confront the mother with the diagnosis in an empathetic yet definite way. She initially could not believe my conclusion. The patient and her mother agreed to keep a skin diary.

Table 23.2 Signs of bullous dermatitis artefacta

Type	Example
Bizar or regular distribution of lesions	Bullae at regular distance (like wallpaper), symmetrical, on arms
Does not fit in known disease	Solitary blister without primary erythema
Medical shopping	Visited several dermatologists including a rheumatologist
'La belle indifference'	Looking untouched while presenting with several painful erosions
Improves under zinc oxide plaster	Healed lesions on lower legs, except at edge of the plaster

Table 23.3 Level of blistering in artificial bullae

Level of blistering	Trauma
Intracorneal	Plucking
Subcorneal of granular	Rubbing
Intrabasal or intraspinal	Electric
Subepidermal	Suction, thermal, acids
Deep cutaneous	Alkalines

Specific Diagnostics

Histopathology of the edge of a blister may reveal the factitious nature. The level of blistering depends on the type of trauma (Table 23.2 and Table 23.3).

The weakest spot (locus minoris resistence) in the skin may divert due to skin disease. For instance, repeated friction by handling a gardening tool (Fig. 23.2) results in a *physiological* friction blister in the granular layer (interface between living and dead epidermis, Figs. 23.3 and 23.4). However, in patients with hereditary epidermolysis bullosa the *pathological* friction blister is intrabasal or subepidermal at the site of the affected adhesion molecule.

Fig. 23.2 Physiological friction blister in normal individual due to repeated trauma with shovel during gardening

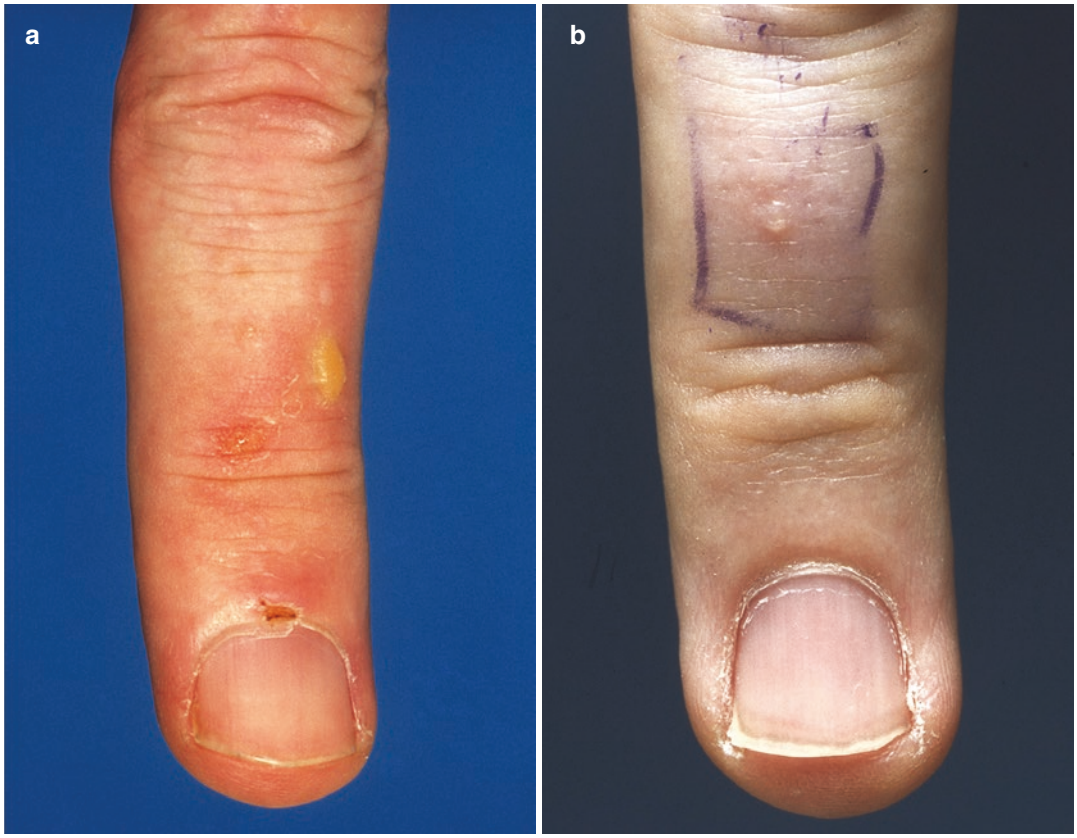

Fig. 23.3 (**a**) Vesicle on the digit in a patient with factitious disorder. (**b**) Patient in (**a**) was able to induce a vesicle in 30 s on the digit of his doctor by friction with his thumbnail (reprinted with permission Ned. Tijdschrift Geneeskd. 2000; 144(31): 1465–9)

Treatment Tricks

Narrow Escape

Tell your patient which diagnoses have been excluded with certainty. Take skin complains seriously and treat symptomatically. Build a safe environment.

First step in treatment is "narrow escape" thus avoiding loss of face of the patient [2]. Create a narrow escape by giving the patient the feeling you know that it self-inflicted, but never directly question it, nor accuse the patient. For instance, at first visit I told a patient during physical examination that I have seen this before, and it remarkably looks like a burn blister. At the end of the consultation I promise the patient to tell what it is at next visit after finishing all examinations. In the meanwhile I ask the patient to keep a dairy of new blisters. Keeping a diary provides extra attention. At second visit the lesions may have cleared. I have heard because the patient stopped drinking coffee, took vitamin C, or confessed to mother when brought to bed. Show happiness and agree with the conclusion of the patient. If the problem persists then introduce the psychiatrist.

The first line management of DA is narrow escape, the second is confrontation

Dual Approach

DA that is not responding to narrow-escape is generally managed by dual (or holistic) approach by dermatologist as the skin expert and the psy-

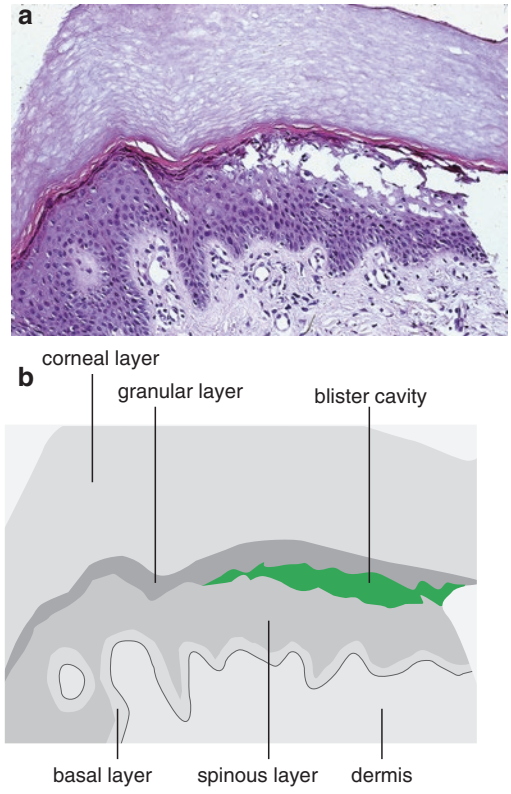

Fig. 23.4 (**a**) Histopathology of physiological friction blister in patient with dermatitis artefacta reveals split level beneath granular layer. (**b**) Diagram depicting (**a**). (Reprinted with permission Ned. Tijdschrift Geneeskd. 2000; 144(31): 1465–9)

chiatrist/psychologist for mental exploration. Offer psychological help by explaining that such as chronic skin disorder will have serious impact on the patient's mental well-being. The psychiatrist may be introduced after the second visit, or be present from the start in special clinics for psychodermatology.

The aim is not to elicit a confession. Patient sins against the ground rule, that he or she is dedicated to be cured. The dual approach also protects the dermatologist from incompetent feelings, elicited by patient demands when relapse occurs. Aggressive emotions in physician may lead to aim of unmasking the patient. Be conscious of this countertransference.

At some stage, in refractory cases, there is no other option than to confront the patient with the self-inflicted nature of the skin problem. This should be done without moral judgment preferable by or in the presence of the psychiatrist. If the patient-doctor relation developed in trust, the patient will not walk away, and let her lesions be treated symptomatically. After all, they also deserve compassion as sufferers of a chronic skin disorder.

Case Study: Part 3

The parents supported her in keeping a skin diary. They noticed repeated rubbing of the face. One night before bed the daughter confessed to her mother that she was nervous at school before math and then rubbed her face. The mother suggested to take a different doll to school every week, and every time she felt nervous cuddle the doll instead of rubbing her face. Complete remission was reached! The doll was the narrow escape introduced by the mother. Other scholars now also took a doll to school to desensitize themselves when nervous. As follow-up, I advised consultation by a pediatric psychiatrist to screen for anxiety disorders in her child.

Review Questions

1. What is most typical of the distribution of DA lesions?
   a. multiple
   b. asymmetrical
   c. trunk
   d. regular
2. The blister level of a thermal blister is
   a. subgranular layer
   b. spinous
   c. intrabasal
   d. subepidermal
3. First approach to a patient with DA is
   a. confrontation
   b. supportive empathy and serious investigation
   c. narrow escape
   d. referral to psychiatrist
4. What examples are NOT a narrow escape

a. starting a food supplement
b. stopping certain food
c. zinc plaster
d. placebo

Answers

1. d.
2. c.
3. b.
4. c.

On the Web

Wikipedia: http://en.wikipedia.org/wiki/Factitious_disorder

Psycho-dermatology, British Association of Dermatology: http://www.bad.org.uk/healthcare-professionals/clinical-services/service-standards/psycho-dermatology

Merck Manuals: http://www.merckmanuals.com/professional/psychiatric_disorders/somatic_symptom_and_related_disorders/factitious_disorder_imposed_on_self.html

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Additional Reading

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