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Lichen striatus in Adults or 'Adult Blaschkitis'?

There Is No Need for a New Naming

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Key Words
Adult blaschkitis · Lichen striatus · Linear inflammatory dermatosis

Abstract
Background: Lichen striatus (LS) is a well-known acquired linear inflammatory dermatosis. In 1990, Grosshans and Marot introduced the term 'adult blaschkitis' (AB), describing an eruption similar to LS occurring in an adult (adult LS). Does there really exist a new entity or a need for a new naming? ***Objective and Methods:*** Two new cases of adult LS are described and the data from 16 earlier cases (12 AB and 4 adult LS) are reviewed. ***Results:*** The analysis of 18 adult patients with an acquired inflammatory blaschkolinear eruption reveals that females are affected two times as frequently as males. The mean age at onset is 44 years, the mean duration until spontaneous cure 8.7 months. Relapses occur in 27.7%. In 78%, the eruption is localized on the trunk, in 55% on the arms and in 50% on the legs. Multilinearity is found in 100% if the eruption is on the trunk, and 61.5% if it is on the limbs. Neither clinical nor morphological differences exist between AB and adult LS. ***Conclusion:*** There are no convincing characteristics which justify creating a new name or even a new entity. AB may be the same as LS, a well-known acquired linear inflammatory dermatosis, which – as has been shown now – does not occur so rarely in adults. However, the etiology of this entity remains obscure.

Introduction

Lichen striatus (LS) is a self-limiting, acquired linear inflammatory dermatosis that occurs most often during childhood and is only seldom seen in adults [1]. It usually manifests itself on the extremities as either a continuous or an interrupted, single or multilinear band, composed of erythematous, sometimes scaly papules. 'Zosteriform' patterns have been recorded. In rare cases, it may also appear on the trunk [2] and the face [3]. The eruption is asymptomatic, has a sudden onset and usually involutes within 1 year [1]. In 1991, Taieb et al. [4] demonstrated that LS follows the lines of Blaschko.

In 1990, Grosshans and Marot [5] described a 38-year-old man with an acquired relapsing inflammatory linear eruption, appearing as slightly pruritic erythematous papulovesicles, following the lines of Blaschko, and showing a nonspecific spongiotic (eczematous) dermatitis on histological examination. The fact that this eruption, with one relapse, occurred in an adult, was mainly localized on the trunk, showed multiple lines and disappeared within less than 2 months caused them to introduce the term 'adult blaschkitis' (AB) for this rare observation. Grosshans and Marot [5] proposed that the rare observations of LS and linear eczema occurring in adults and following the lines of Blaschko could also be adjusted to this new term. Since then, other case reports have followed [6–20]. Even if some authors are trying to find a difference between AB and LS [14, 19], the discussion remains controversial. In trying to unite these two entities, the term 'Blaschko linear acquired inflammatory skin eruption' (BLAISE) has been created [4]. In 1994, Megahed et al. [9] proposed the term 'acquired relapsing self-healing Blaschko dermatitis' to differentiate this condition from LS. In 1996, Lee et al. [14] tried to simplify this long and complex name by proposing 'acquired Blaschko dermatitis' still intending to differentiate it from the adult onset of LS.

The personal observation of two women with an acquired inflammatory eruption following the lines of Blaschko gives reason to review the recent literature [5–20] and to discuss once more whether there really exist differences between AB and LS occurring in adults and then to address the question whether there is a need for a new naming.

Case Reports

Case 1

A 37-year-old mother and housewife noticed an asymptomatic, nonpruritic rash following her third pregnancy. Topical treatment with a cream containing methylprednisolone aceponate induced an almost complete clearance except for the remaining of some slightly hyperpigmented patches. Beside a well-known atopic diathesis with hayfever and marked neurodermatitis in childhood, which by now have almost disappeared (except some exacerbation during the pregnancies), her personal medical history was absolutely uneventful. She gave birth to

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3 healthy children in 1992, 1996 and July 1999 after undergoing normal pregnancies. Following her last delivery, the patient has had an intrauterine device containing 52 mg levonorgestrel. She denied any other drug intake. Physical examination in December 2001 revealed barely visible discrete hyperpigmented streaks in a 'zosteriform' pattern on the lower part of her right hemithorax. The patient was advised to stop any treatment and 5 weeks later, a reddish, papular, scaly dermatosis in linear arrangement following the Blaschko lines, strictly limited to the right side of her abdomen (fig. 1) could be noted. The same eruption, but in a discrete, multilinear pattern, could also be seen on the anterior aspect of her right lower leg. On histological examination, an inflammatory infiltrate of lymphocytes and few histiocytes, showing some exocytosis, was present in the upper dermis, just beneath the epidermis, which showed moderate acanthosis and focal parakeratosis. Direct immunofluorescence was negative. The diagnosis of AB was assumed. Again 5 weeks later, the clinical picture had changed in a way that the scaly character had almost disappeared, and there were shiny lichenoid coalescing papules measuring 1–3 mm in diameter. On histological examination, a fairly heavy infiltrate, most of them CD3 positive lymphocytes and histiocytes, lied in close approximation to the epidermis, which itself was flattened, showed discrete hydropic degeneration, few dyskeratotic keratinocytes, an almost disappeared granular layer and elongated rete ridges. A lack of violaceous color and Wickham's striae on the papules and, on histological examination, a lack of hyperkeratosis, focal hypergranulosis and irregular acanthosis [21] helped exclude the diagnosis of lichen planus. Even if the picture histologically resembled that of lichen nitidus (except the lack of epitheloid cells or Langhans giant cells), the clinical aspect did not fit in it. So, the diagnosis of LS in an adult was made. Laboratory test showed a normal hematogram. C-reactive protein and erythrocyte sedimentation rate were normal. There were no detectable autoantibodies (ANA, anti-DNA).

Case 2

In May 1991, a 46-year-old woman was admitted because she noticed a slightly pruritic rash, which had begun 6 months ago on the medial aspect of her left heel and was slowly spreading in a linear fashion, upwards the inner side of her calf and thigh up to her left labium majus and downwards to the basis of her hallux. The eruption consisted of erythematous papules which became somewhat pruritic and hyperkeratotic near the ankle and the upper thigh. On histological examination, an inflammatory infiltrate was seen around the subpapillary vessels, extending into some of the papillae. The epidermis showed exocytosis and spongiosis, few dyskeratotic keratinocytes, slight hyperkeratosis, and no acanthosis or parakeratosis. The diagnosis of LS in an adult was made. A cream containing betamethasone was prescribed against the itch. Within 2 months, the eruption disappeared without any sequel. Her personal medical history revealed three uneventful pregnancies. Beside an atopic diathesis with hayfever and asthma, induced by an allergy to grass and rye pollens, she suffered an episode with recurrent angioedema and urticaria caused by an aspirin intolerance in 1989.

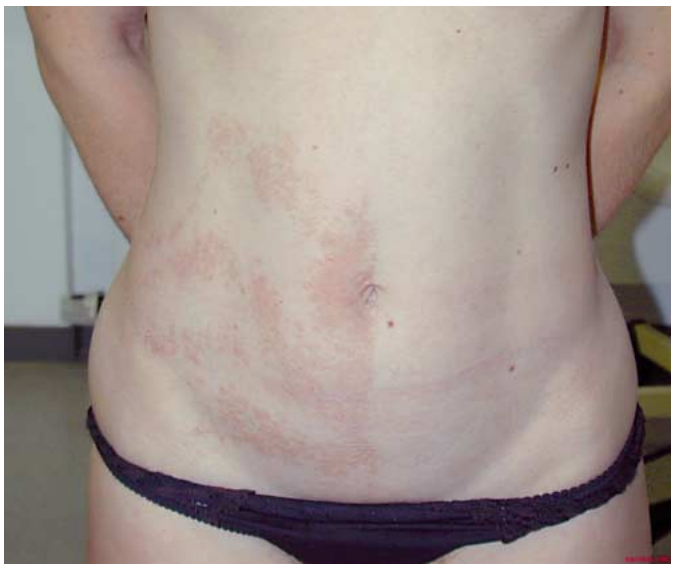

Fig. 1. Case 1, showing a multilinear eruption following the lines of Blaschko on the right side of her abdomen.

Discussion

Congenital, nevoid, acquired skin diseases, but also acquired inflammatory dermatosis may follow the lines of Blaschko. They are thought to be caused by genetic mosaicism, resulting from lyonization in X-linked disorders, postzygotic somatic mutations in sporadic conditions and gametic half-chromatid mutations [22]. Another genetic concept, loss of heterozygosity in human skin, may play a role not only in cutaneous malignant growth but also in the development of benign skin disorders [23]. Lipsker et al. [24] for the first time demonstrated that an acquired inflammatory dermatosis in blaschkolinear distribution does indeed show genetic mosaicism. LS occurs most often in children. If it is observed in adults, then it shows close similarity with AB, a relapsing inflammatory dermatosis following the lines of Blaschko, first described by Grosshans and Marot in 1990 [5]. Since then, 15 similar cases have been reported [6–20]. Eleven of these reports claim to present another example of this new entity [6–10, 12–14, 16, 17, 19], whereas 4 report the rare variant of adult LS [11, 15, 18, 20]. All the reports discuss the difficulty of the distinction between these two diseases. While some [6, 16, 20] presume that they merely represent either end of a spectrum, others [14, 19] suggest criteria for a clear differentiation between the two disease entities (table 1). Reiter et al. [18] presume that AB is nothing else than a variant of LS and Tomasini et al. [11] think that the term AB is just a new name for adult LS.

The review of the literature [5–20], together with the 2 new cases presented here show that there hardly exist any differences between AB and adult LS concerning the mean age of patients, mean duration, localization and multilinearity of the eruption. Even if there are some differences between AB and adult LS comparing sex ratio as well as percentage of relapses, the two groups are too small to favor a significant difference (ta-

ble 2, first and second column). True bilateral lesions are only found in 1 patient with AB [19]. One patient shows them symmetrically on the penis [10]. All who claim a clear difference between AB and LS (table 1) emphasize that the former is characterized by papulovesicles and histological spongiotic changes, whereas LS predominantly shows papules and histological lichenoid changes [14, 19, 25]. Spongiosis (intercellular edema) and exocytosis of lymphocytes are mentioned in all reported cases of adult LS but only in 9 (spongiosis) and 8 (exocytosis) of 12 cases with AB (table 3, first and second column). It is of interest that only 3 cases of AB showed intraepidermal vesicles [7, 9, 16], whereas in two thirds, a papulovesicular eruption [5–9, 12, 14, 17] is described! The development of micro- and – finally visible – macrovesicles depends on the degree of spongiosis and intracellular edema [26], which again might reflect the intensity of the inflammatory reaction and the acuteness of the eruption. Indeed, Grosshans and Marot [5] have already mentioned in their first reported case that the relapse was characterized by 'papules fermes' (firm papules) and not by papulovesicles. All except 1 [8; no histopathological examination] case reports describe an inflammatory focal or lichenoid infiltrate composed mainly of mononuclear cells in the upper dermis. In 50% of AB and adult LS the infiltrate is also seen around vessels and adnexa in the deeper dermis. In summary, all these histological results (table 3, last column) are in concordance with studies describing the histological and immunohistochemical characteristics of LS [27, 28]. So, it is possible to conclude that there exist neither clinical nor morphological differences between AB and LS occurring in adults. That means, the proposed characteristics (table 1), which should enable to differentiate AB from adult LS, are not robust enough. In the discussions regarding the pathogenesis of AB or adult LS, it is of interest that in these reports the cited articles always concern LS. The main hypothesis is that, beside genetic factors [29], acquired stimuli, like a viral infection [3] or a trauma, induce loss of immunotolerance resulting in a T-cell-mediated inflammatory reaction against keratinocytes, which themselves show genetic mosaicism and are arranged along the lines of Blaschko [11, 14, 16–18]. Also the hypothesis of a somatic mutation of a keratinocytic clone, which could induce an autoimmune response of the host [27], similar to the graft-versus-host reaction, is cited [15, 17, 18].

Table 1. Distinguishing factors between AB and LS [14, 19, 25]

AB	LS	References
Rare	More common	25
Adults	Children (mean age 3 years)	25, 19, 14
Trunk predominantly	Limbs predominantly	25, 19
Papules and vesicles	Papules, rarely vesicles	25, 19, 14
Multiple lines	One or few lines	25
Uni- or bilateral lesions	Unilateral lesions	25, 19
Rapid resolution (less than 2 months)	Slow resolution (3 months to 2 years)	25, 19, 14
Relapses	Rare relapses	25, 19, 14
No sequelae	Transient hypopigmentation	25
Spongiotic histological changes	Spongiotic/lichenoid changes	25, 19, 14

Table 2. Clinical characteristics

	AB (12 patients)	Adult LS (6 patients)	AB and adult LS (18 patients)
Sex	5 ♀ 7♂	6 ♀	11 ♀ 7♂
Age at onset, years	ø 42.0; 24–70	ø 47.3; 36–64	ø 44; 24–70
Total duration, months	ø 8.5; 1–39	ø 9.2; 1–32	ø 8.7; 1–39
Relapse	4 (33.3)	1 (16.6)	5 (27.7)
Location			
Trunk	9 (75.0)	5 (83.0)	14 (78.0)
Arm	8 (66.6)	2 (33.3)	10 (55.5)
Leg	5 (41.7)	4 (66.7)	9 (50.0)
Multilinearity			
Trunk	(100.0)	(100.0)	(100.0)
Limbs	(62.5)	(60.0)	(61.5)

Figures in parentheses are percentages.

Table 3. Histological characteristics

	AB (12 patients)	Adult LS (6 patients)	AB and adult LS (18 patients)
Spongiosis	9	6	15
Exocytosis	8	6	14
Dyskeratotic keratinocytes	5	5	10
Focal parakeratosis	5	3	8
Acanthosis discrete	5	3	8
moderate	3	0	3
Hyperkeratosis	1	3	4
Mononuclear infiltrate			
Upper dermis (focal/lichenoid)	11	6	17
Mid-dermis (adnexa/vessels)	6	3	9

In 1988, Burton et al. [2] wrote in their *Textbook of Dermatology* that LS may rarely be seen in adults. The lesions of LS occur commonly on the limbs, but may develop on the trunk. Bilateral involvement is exceptional. The eruption is characterized by one or more parallel linear band, 'zosteriform' patterns are possible. In 1974, Charles et al. [1] noted that females are affected two or three times as frequently as males and that the eruption of LS usually involutes within 1 year. This longstanding description of LS fits exactly in the results obtained by the review of the 18 cases presented here with an acquired inflammatory dermatosis occurring in adults and following the lines of Blaschko (table 2, last column). The only latest findings we can add are that AB/LS always follow the lines of Blaschko [4], that they do show genetic mosaicism [24] and, in rare cases, may relapse. But these three new findings are not sufficient enough to create a new name or even a new entity, especially since the origin of AB and/or LS remains unknown.

'-itis', a common ending for inflammatory processes in medicine, can hardly be reserved for a singular inflammatory reaction occurring along the lines of Blaschko, as it is done with the term 'adult blaschkitis'. Also other inflammatory dermatosis like lichen planus [30, 31], lichen nitidus [32, 33], lupus erythematosus [13] and even psoriasis [22] may follow the lines of Blaschko representing an '-itis' in rare cases. But it is of special interest that in these cases, a well-known clinical entity shows this rare pattern, whereas in LS/AB, we still miss any knowledge of a corresponding entity. In 1986, Toda et al. [34] stated that 22 of 26 patients with LS had a positive history of atopic disorders. Taieb et al. [4] noted that one third of their reported cases with LS had associated cutaneous findings consistent with mild atopic dermatitis. Also the two new presented cases have a positive history of atopic disorders. Unfortunately, none of the reviewed reports [5–20] mentions anything about possible associated atopic disorders. Even if it is conceivable, it remains hypothetical whether LS represents a linear pattern of neurodermatitis. Furthermore, the debate is still going on whether LS and linear lichen ruber represent opposite ends of the same spectrum [15, 35].

In conclusion, acquired inflammatory dermatosis following the lines of Blaschko, occurring in adults and resolving without any treatment, may represent LS. The review of the data from 16 earlier cases and the 2 new presented cases do not justify new names as there are, i.e. 'adult blaschkitis' [5], 'acquired relapsing self-healing Blaschko dermatitis' [9] or 'acquired Blaschko dermatitis' [14]. There still remains the difficulty in differentiating LS from other acquired inflammatory dermatosis, rarely following the lines of Blaschko, as lichen planus [30, 31], lichen nitidus [32, 33], lupus erythematosus [13] and psoriasis [22].

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