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Lichen striatus: acquired dermatitis on a silent nevus.

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Summary Lichen striatus (LS) is an acquired, asymptomatic dermatitis, usually distributed monolaterally on a skin segment, which heals spontaneously in months; the most affected age group is that of the kindergarten. Despite its banality LS always comes to the doctor due to the unusual distribution of the lesions. This report is a retrospective analysis of the clinical data of 160 cases of LS in children aged less than 13 years, 67 males and 93 females. Its epidemiological, clinical and pathogenetic data are discussed.

Key words Lichen striatus, Blaschko, virus.

Lichen striatus (LS) is an acquired, asymptomatic dermatitis, characterized by small papules usually distributed monolaterally on a skin segment, which heals spontaneously in months; the most affected age group is that of the kindergarten. Despite its banality LS always comes to the doctor due to the unusual distribution of the lesions.

This report is a retrospective analysis of the clinical data of 160 cases of LS in children aged less than 13 years. It is aimed at spreading awareness of a disease that any doctor can follow reassuring the parents about its benign course.

Material and results

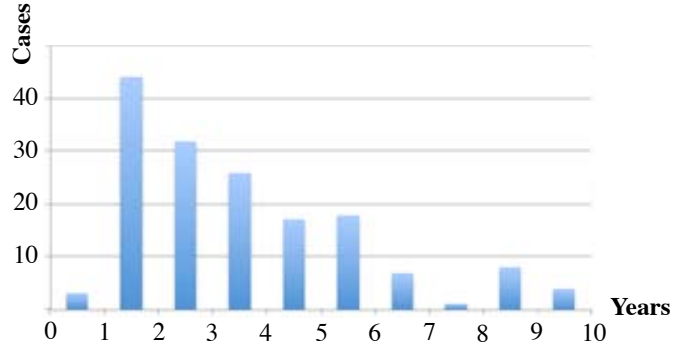
160 Caucasian children aged less than 13 years consecutively observed in our outpatient Pediatric Dermatology clinic between 1975 and 2013 entered the study.

The epidemiological data (age, sex, season of onset of the disease), history and subjective symptoms, physical examination (site involved, skin lesions and their outcomes) and therapy of these children were reviewed.

Of the 160 children 93 were males and 67 females. The age of the child at the time of the visit (Fig. 3) practically coincided with the chronological age because LS, due to its unusual distribution, is observed by the physician within a few days or a few weeks after the onset: in particular 3 children were aged less than 1 year, 44 between 1 and 2 years, 32 between 2 and 3, 26 between 3 and 4, 17 between 4 and 5, 18 between 5 and 6, 7 between 6 and 7, 1 between 7 and 8, 8 between 8 and 9, 4 between 9 and 10.

LS was often preceded by an inflammatory episode of the respiratory tract. In cases where the

Table 1: age of onset of lichen striatus.



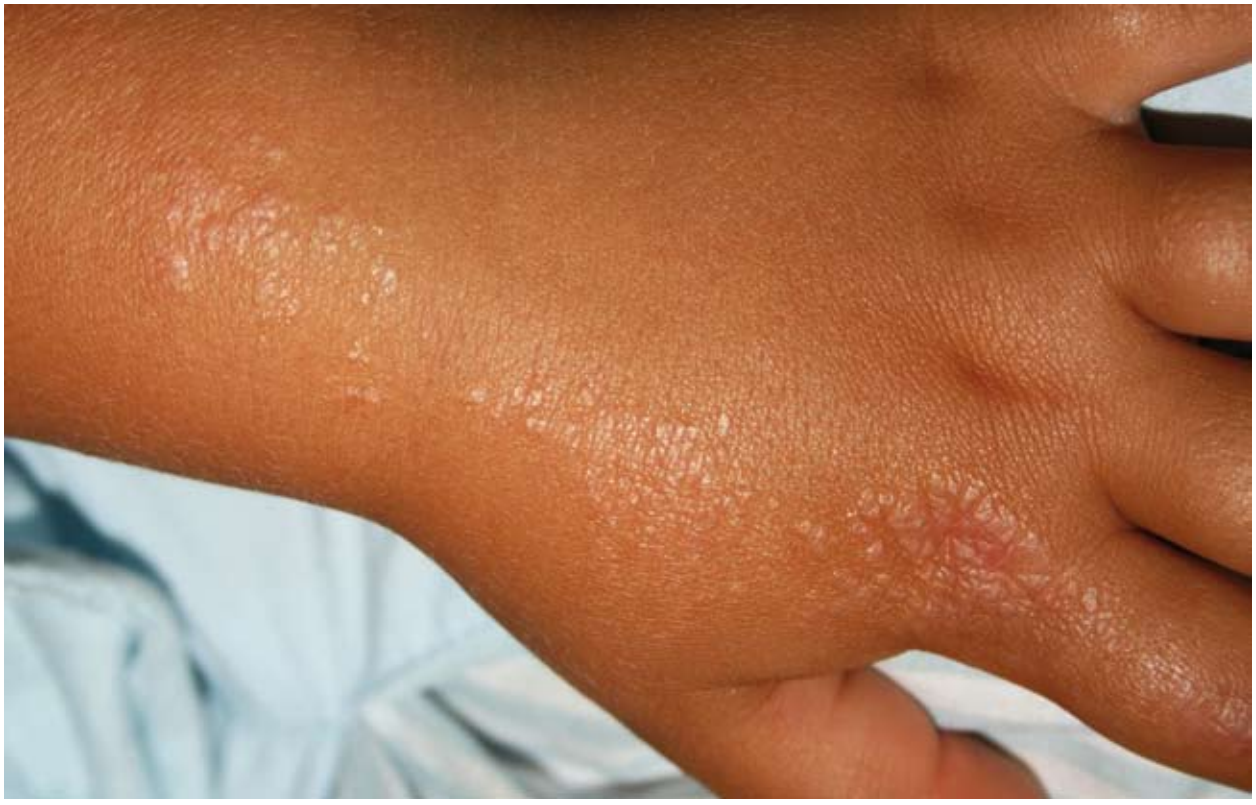


Fig. 1: Lichen striatus: 0.5-1mm in size papules monolaterally distributed on a skin segment.

physical examination at the diagnosis highlighted only hypomelanic residua that due to their shape, size and distribution led to the diagnosis of LS, the history put in evidence the previous presence of reddish papules only in 27/51 cases. In all cases the itching was absent or moderate. The inflammatory phase, which was not always clinically evident, typically lasted 2-3 weeks, but in 3 cases more than 6 months; the hypomelanic residua usually subsided within a year. In our study, we observed no recurrence of the lesions.

The affected site was the head-neck in 31 cases, trunk in 27, upper limbs in 49 and lower limbs in 53 cases (Fig. 1, 2, 3, 4, 5). In 4 cases more than 1 segment was involved: upper limb and trunk in 2 cases, lower limb and trunk in the other 2. In 4 cases the involvement of a hand was associated with nail involvement (Fig. 7).

The physical examination revealed pink or red 0.5-1mm papules monolaterally distributed on a single skin segment in 97 cases; in 51 cases there were only hypomelanic residua (Fig. 2) of the same size of papules; in 12 cases there were both papules and hypomelanic residua. Both papules and hypomelanic residua were unilateral and followed the lines of Blaschko; only in one case the papules on the back also affected the contralateral segment (Fig. 8).

In 153 cases no therapy was prescribed; in 7 cases with papular lesions in visible sites topical corticosteroids were given until improvement of the lesions.

Discussion

Lichen striatus (LS) arrives to the physician a short time after its onset. Characteristically the recently acquired lesions are distributed along the lines of Blaschko; namely they run longitudinally along the limbs, form spirals on the sides and "V" in the vertebral region (Fig. 5, 8). Sometimes the lesions are distributed along two (Fig. 3) or more parallel lines, separated from each other by

skin of normal appearance. The lesions consist of 0.5-1 mm in diameter papules with flat surface or pointed, more or less red, confluent when the inflammation is intense, sometimes scaly on the hands (Fig. 6) and feet where the horny layer is thicker, and exceptionally vesicular (8).

After weeks or months the red papules are substituted by hypomelanic residua, which sometimes are the only sign of the disease from its onset: the hypomelanic residua have the same size and the same distribution of papules. Rarely, the inflammatory active phase of the disease can last for years (2).

The lesions, when affecting the nail matrix, are responsible for alterations of the nail lamina (Fig. 7). The latter, like all the lesions of this site, take much longer to regress compared to skin lesions; exceptionally the scalp can be affected with transient hair loss (10).

Once regressed, the lesions do not recur any more, thus behaving like a viral rash that leaves permanent immunity. The differential diagnosis of LS should be done with all the skin disorders distributed along the lines of Blaschko. Very important is the question "how long", which allows to rule out inherited skin disorders, such as incontinentia pigmenti (Fig. 11) and clinically obvious nevus lesions such as epidermal nevus (Fig. 14); among the nevus lesions inflammatory linear verrucous epidermal nevus mostly looks like LS (Fig. 12). However, it is more precocious, intensely itching and persists indefinitely.

The differentiation from other acquired diseases distributed along the lines of Blaschko is less easy, especially when there are no milder lesions of the same disease bilaterally distributed on the entire skin surface. In the latter case physicians should search for the clinical characteristics of

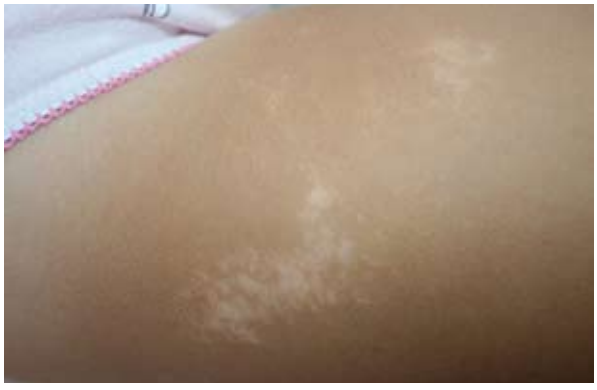


Fig. 2

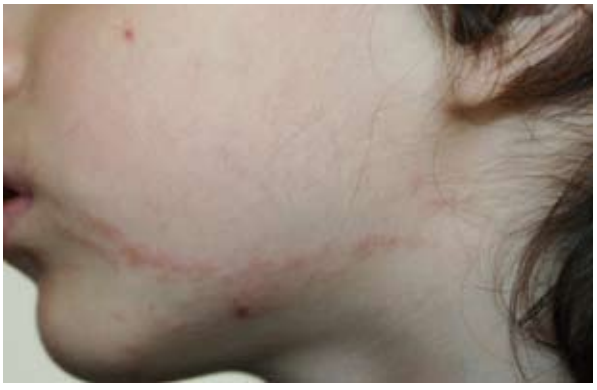


Fig. 3

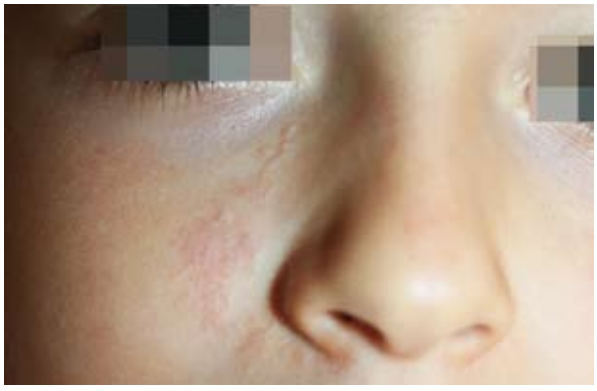


Fig. 4

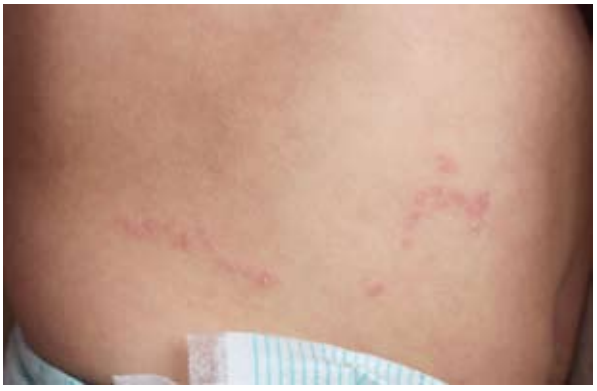


Fig. 5

Fig. 2, 3, 4, 5: Hypomelanic residua of lichen striatus (LS) of the thigh. In Fig. 3 LS of the mandibular region: you can see the two parallel streaks of pathological skin separated by a streak of normal skin. In Fig. 4 LS of the right paranasal region. In Fig. 5 LS of the right side: you can see the "S" shape.

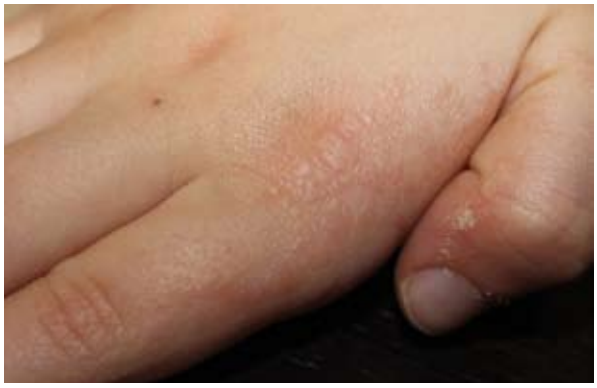


Fig. 6

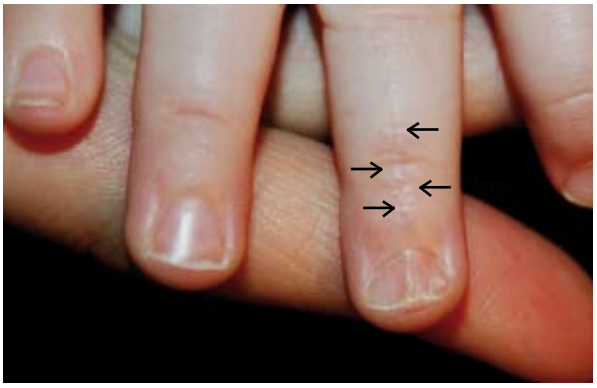


Fig. 7

Fig. 6, 7: LS of the first and second right finger: the papules are associated with scales where the horny layer is thicker on the ulnar surface of the thumb. In Fig. 7 LS of the third finger (arrows) with involvement of the nail matrix and consequent longitudinal ridge of the lamina.

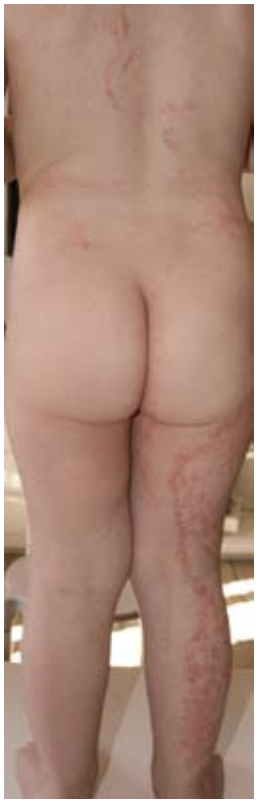


Fig. 8

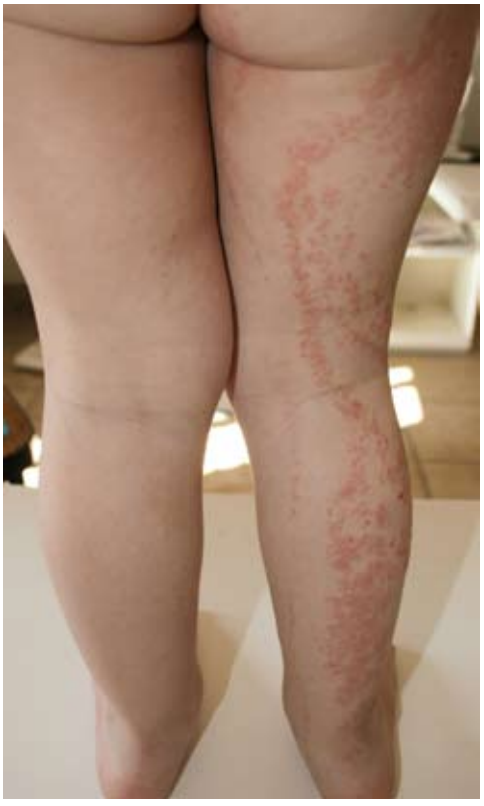


Fig. 9

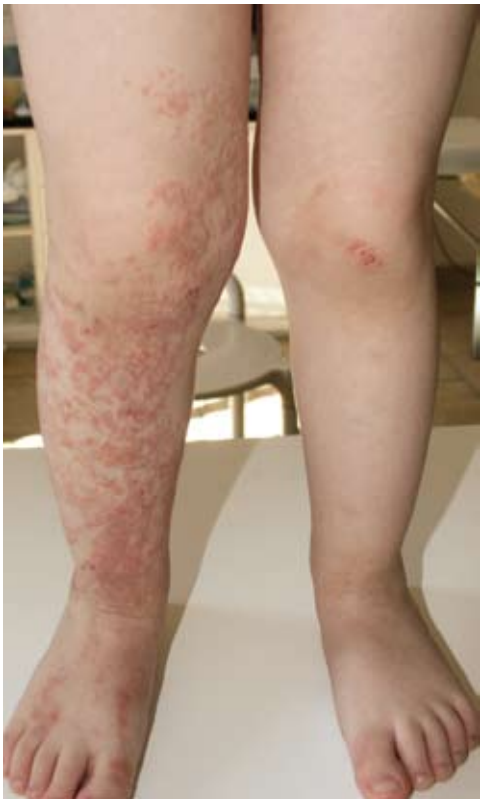


Fig. 10

Fig. 8, 9, 10: Right unius lateris LS encroaching on the left side level with the back. On the thigh (Fig. 9) you can see the bend with medial convexity of the Blaschko lines.

the acquired disease: linear psoriasis (Fig. 15) will be differentiated thanks to the more important scaly component, for its longer persistence, for the response to the sun and topical immunosuppressive drugs; linear vitiligo (Fig. 16) thanks to the reddening after sun exposure, the possible presence of follicular repigmentation and the variation of its extension with time; lichen planus thanks to the presence of Wickham striae well visible dermoscopically (12) and lichen nitidus

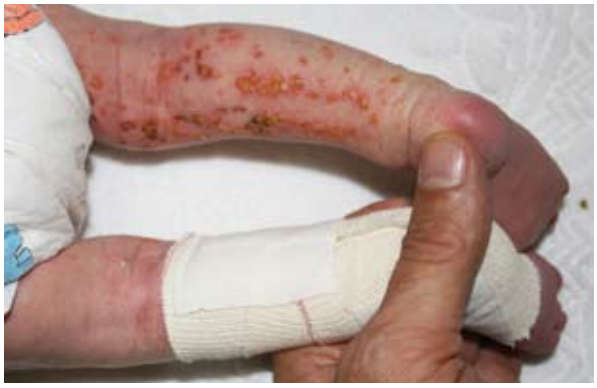


Fig. 11: Blaschko-linear bullous lesions of incontinentia pigmenti at birth.

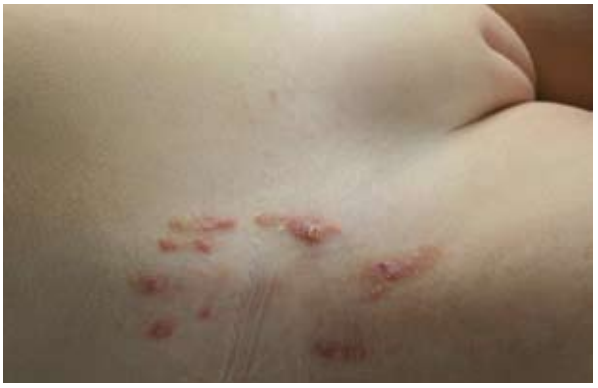


Fig. 12: Inflammatory linear verrucous epidermal nevus present since birth.

thanks to its less inflammatory papules with typical histology.

LS seems to be due to an environmental agent which makes its action only on an abnormal keratinocyte population susceptible to that agent. The hypothesis of a pre-existing population of mutated keratinocytes (11) is based on the particular distribution of the lesions: linear, sometimes in parallel streaks separated by streaks of healthy skin. This distribution implies the existence of a cutaneous mosaicism, i.e. of two cell populations, one normal and the other pathological. All diseases - inherited, nevus, acquired - with this type of distribution are due to a mutation, present in the first cell or zygote - hereditary diseases - or occurred in a primordial, post-zygotic cell during

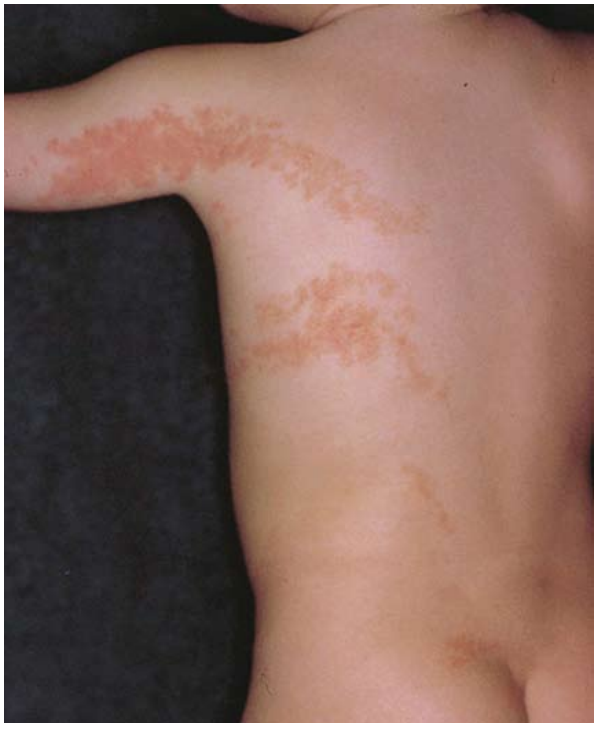


Fig. 13

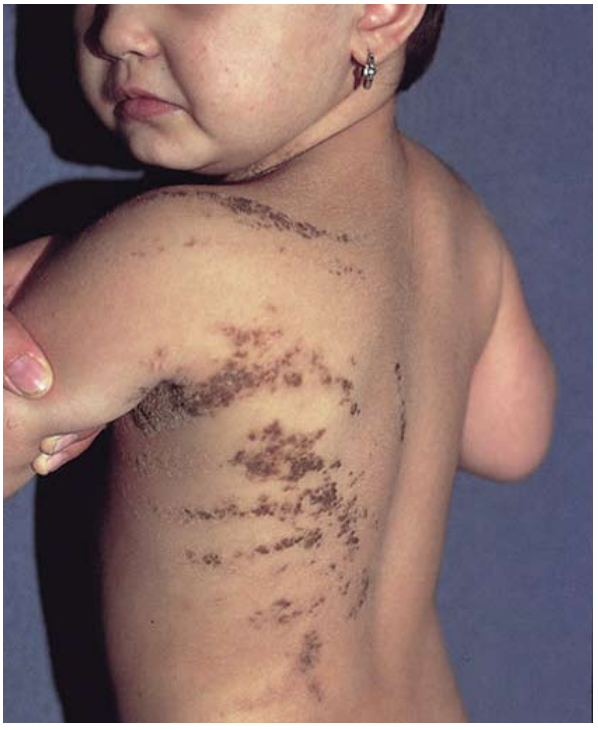


Fig. 14

Fig. 13, 14: Lichen striatus (Fig. 13) and epidermal nevus (Fig. 14) can be distributed in the same way; however, the latter is present since birth.

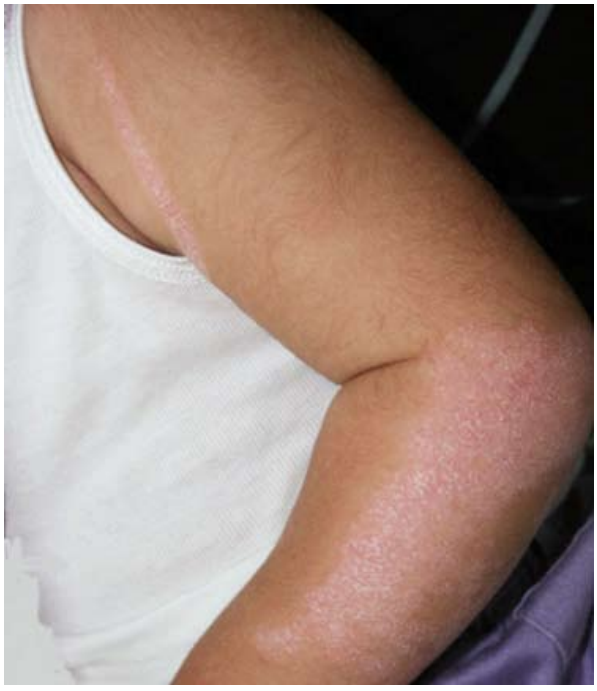


Fig. 15:Blaschko-linear psoriasis of the left upper limb: it is more persistent than LS, regresses during summer and is more scaly.

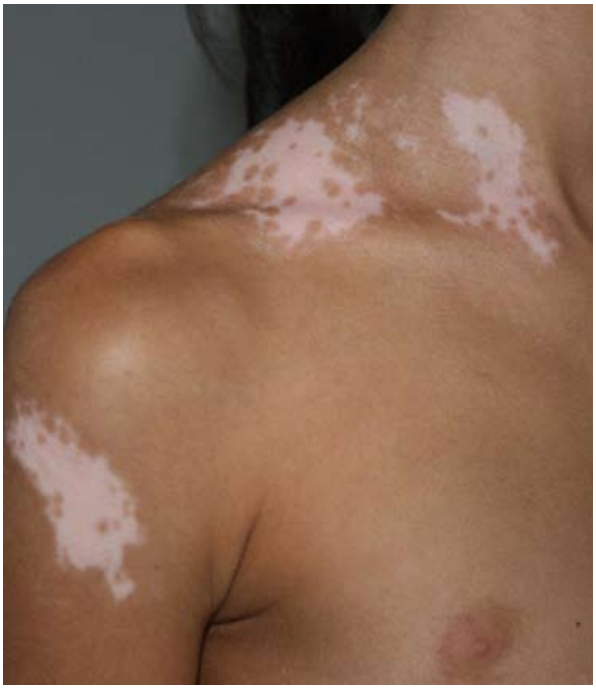


Fig. 16: Segmental vitiligo: it can be differentiated from hypomelanic residua of LS mainly due to the follicular repigmentation.

fetal life - nevus and acquired diseases -. The mutation, when occurring in a primordial skin cell during the fetal life, gives rise to a pathological clone of cells that migrate in the fetus following the Blaschko lines, i.e. the binaries along which run the skin cells, particularly the keratinocytes (6).

The mutation is clinically evident from birth in the nevus, for example, the epidermal nevus (Fig. 14); on the other hand, in the acquired diseases distributed along the lines of Blaschko the mutation, clinically silent at birth, gets visible when an environmental factor, acting on the mutated cells particularly susceptible to that factor, causes inflammation that makes it clinically visible.

Most of the acquired diseases which can be distributed along the lines of Blaschko are autoimmune in nature and the trigger factor is likely different according to whether it is lupus erythematosus or psoriasis or vitiligo or lichen planus or another disease.

In lichen striatus the trigger agent may be a virus (3). This hypothesis is supported by the most frequent age of onset, no recurrence of the lesions, contemporary onset in twins and higher concentration of cases in the spring.

The age of onset of LS is that of kindergarten when viral rashes are common, as evidenced by our series, in which nearly 50% of cases affects the 2nd and 3rd year of life and 87% of cases is included in the age group between 2 and 6 years. However, it is not easy to ascertain the role of previous respiratory infections, given the frequency of these episodes in the kindergarten age when most cases of LS occur.

In our series we did not observe LS recurrence, but there are recurrent cases in the literature (11), especially in adults. Probably, the difference between the cases of the child and those recurrent in the adult (5) is not substantial, implying always a clinically silent nevus and an exogenous causative agent; perhaps the difference consists of a different mutation that predisposes to different environmental agents, even for instance viruses that do not give lifelong immunity such as those of the common childhood rashes.

Also important is the report of contemporary onset of LS in twins (9), which is reminiscent of

a particular susceptibility to a common environmental factor.

Less important seems the most frequent occurrence of LS in the spring, as suggested by some Authors (6): in our series about one third of cases occurs in the spring.

If the viral hypothesis was valid, the virus could act directly on the mutated predisposed cells infecting them and then eliciting an immune protective response able to prevent recurrences (1); or the virus could induce the expression of a new membrane antigen on the mutated cells and thus lead to an autoimmune reaction in the organism: the presence of lymphocytes CD8+ around necrotic keratinocytes (4) would seem to confirm the hypothesis of a cytotoxic process directed against the keratinocytes of the mutated clone (11).

In conclusion, it is useful that the doctor even non-specialist in dermatology, knows the lichen striatus to reassure parents about its benign clinical course and spontaneous regression.

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