

BRIEF REPORT

Superimposed linear atopic dermatitis

Abstract

Superimposed linear atopic dermatitis is rarely manifested in polygenic disease as a counterpart to type 2 segmental mosaicism of monogenic skin diseases. Linear arrangement following Blaschko lines represents more severe disease on a generalized background of atopic dermatitis, perhaps reflecting clonal loss of heterozygosity. Only three cases of superimposed linear atopic dermatitis have been reported; we describe three additional cases.

1 | INTRODUCTION

In a patient with polygenic skin disease, the presence of more severe cutaneous involvement of the same disease showing a mosaic arrangement is called a superimposed segmental manifestation of the skin disease.¹ Examples of this include linear dermatomyositis on a background of disseminated dermatomyositis,² segmental vitiligo in patients with generalized vitiligo,³ linear psoriasis in patients with underlying plaque-type psoriasis,⁴ and linear lichen planopilaris in patients with classical lichen planopilaris.⁵ Superimposed linear atopic dermatitis has been rarely described.^{6,7} We present three patients with persistent, linear atopic eczema superimposed on a background of chronic, generalized atopic eczema.

2 | CASE REPORTS

Three patients, a 4-year-old girl, a 2-year-old girl, and a 15-year-old boy (Table 1, Figures 1 and 2), presented with linear eczematous skin disease in the relapsing course of atopic dermatitis. One patient (Case 3) also had atopic asthma. Examination revealed erythematous, extremely pruritic papules, vesicles, and excoriations following a unilateral linear arrangement. They were located on the right leg (Cases 1 and 2) and the trunk and limbs on the left side (Case 3). All patients also had widespread disseminated involvement with milder lesions (Figures 1B and 2B,C). Initiation of systemic and topical therapy was proposed.

3 | DISCUSSION

In monogenic disorders, there are two types of segmental mosaicism due to a postzygotic mutation. In type 1 segmental mosaicism, the individual presents solely with a segmental manifestation of an autosomal-dominant disease. In type 2 segmental mosaicism, the individual has diffuse, bilaterally arranged skin disease and a more severely involved area of the same disease in a segmental arrangement.^{1,2} Type 2 segmental mosaicism implies a heterozygous germline mutation and loss of heterozygosity (LOH) during early embryonic

TABLE 1 Summary of reported cases of superimposed linear atopic dermatitis (AD)

Reference	Age, years	Sex	Location of segmental lesion	Arrangement	Concomitant relapsing AD	Age at onset of linear AD, years	Age at onset of generalized AD	Evolution	Atopic disease background
Taïeb et al ⁶	8	Female	Left arm from axilla to wrist	Linear, unilateral	+	7	6 mo	Improved after 6 wk	Eczema, house dust mite, cat and dog dander, allergy
Hadlik et al ⁷	36	Male	Left lower abdomen, leg, foot	Linear, unilateral	–	36	Childhood	Chronic, recurrent	Allergic rhinoconjunctivitis
Kawachi et al ⁸	65	Male	Left leg	Linear, unilateral	+	62	45 y	Chronic, recurrent	Chronic prurigo
Case 1	4	Female	Right leg	Linear, unilateral	+	4	6 mo	Unknown	Eczema
Case 2	2	Female	Right leg	Linear, unilateral	+	2	2 y	Improved after 4 wk	Eczema
Case 3	15	Male	Trunk; left arm, leg	Linear, unilateral	+	15	2 mo	Chronic, recurrent (followed for 1 y)	Asthma

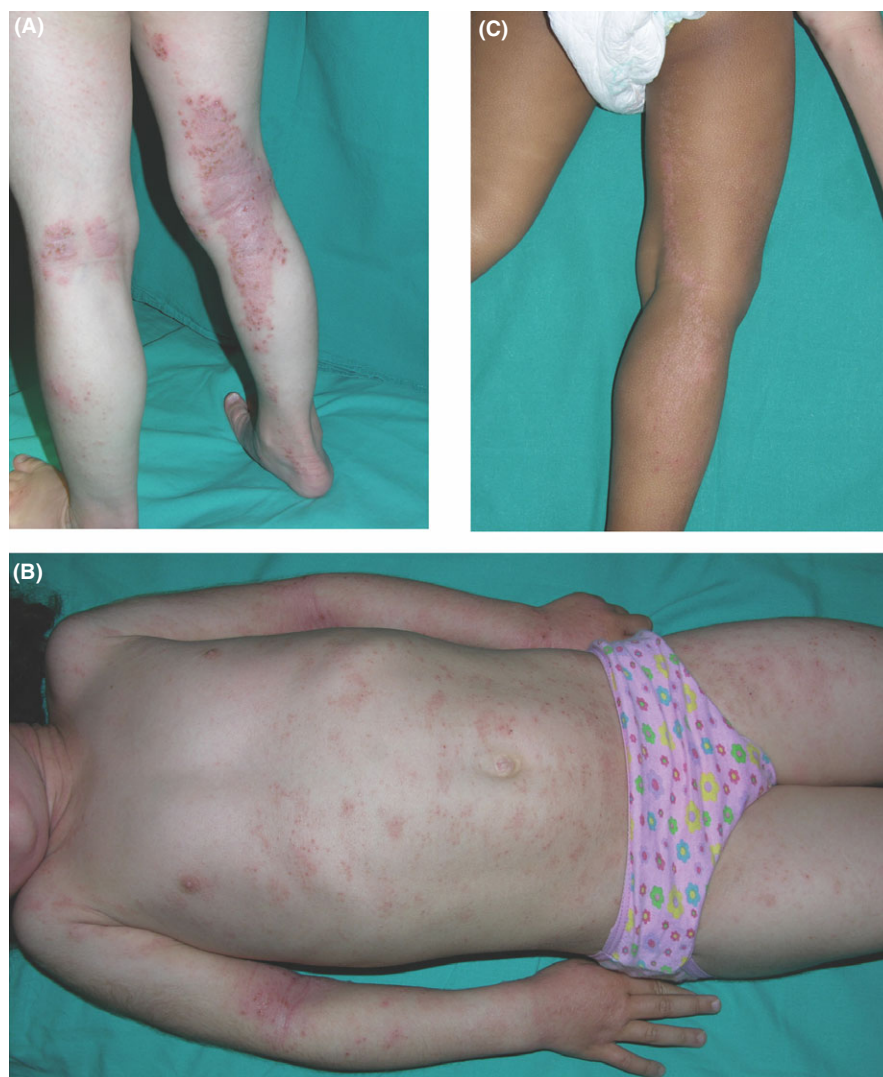


FIGURE 1 Case 1: A, Linear arrangement of lesions in the inner part of her right leg. B, Generalized papules and plaques. Case 2: C, Mild atopic dermatitis and a line of eczema on the back of her right leg

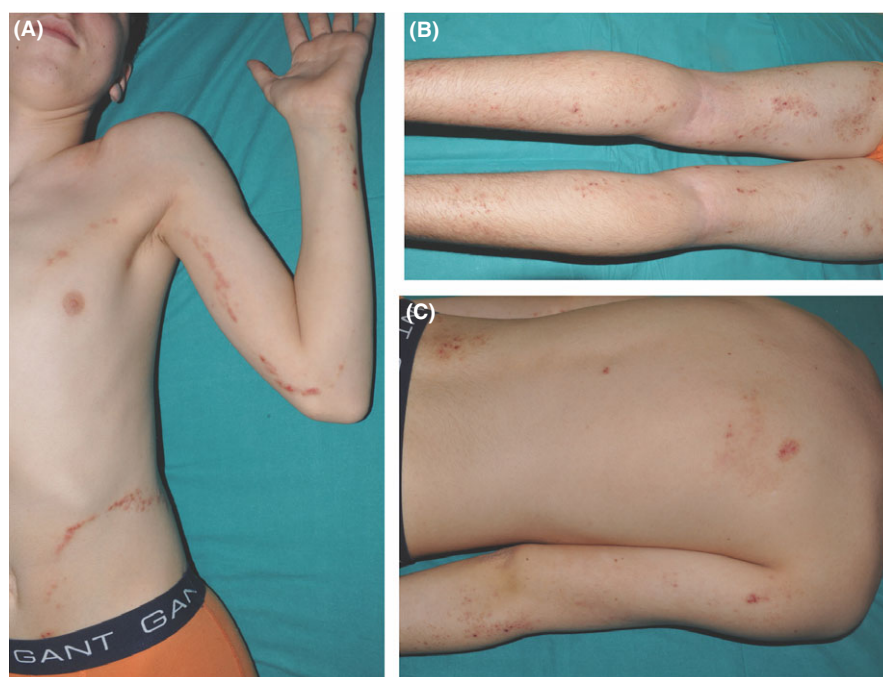


FIGURE 2 Case 3: A, Eczematous lesions following a Blaschko linear pattern affecting the left side of the body trunk and arm. B, C, Diffuse eczema papules and plaques on the back and both legs

development, with loss of the corresponding wild-type allele. Similar skin involvement is known to occur in polygenic skin diseases, in which more severe, segmental lesions can coexist with the nonsegmental skin condition. This phenomenon is known as superimposed segmental manifestation, as a counterpart of type 2 segmental mosaicism of monogenic skin diseases. It has been proposed that, according to the “ $n + 1$ ” rule, in an individual carrying an unknown number of alleles predisposing to a given polygenic disorder, early LOH at one of these gene loci or an early postzygotic new mutation occurring in an additional predisposing gene may render a segmental cutaneous area predisposed to more severe involvement.¹

Only three cases of superimposed linear atopic dermatitis have been reported; we describe three additional cases (Table 1).^{6,8} Linear skin involvement may appear at any age and coincides with disease flares; all six patients had generalized atopic dermatitis^{6,7} or chronic prurigo⁸ before the linear involvement became apparent.

We speculate that this phenomenon may be more common than reported, especially because of the high prevalence of atopic dermatitis, but may have gone unnoticed so far.

Keywords

allergy, atopic dermatitis, genetic diseases/mechanisms

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