

CASE REPORT

A linear lesion in a child with atopic dermatitis: Not a coincidence

Julie Van Gysel¹ | Ramon Grimalt²¹Université Libre de Bruxelles, Brussels, Belgium²Facultat de Medicina i Ciències de la Salut, Universitat Internacional de Catalunya, Barcelona, Spain**Correspondence**Ramon Grimalt, Facultat de Medicina i Ciències de la Salut, Universitat Internacional de Catalunya, Josep Trueta s/n 08195 Sant Cugat del Vallès, Barcelona, Spain.
Email: rgrimalt@uic.es**Abstract**

A blaschkolinear lesion in a patient with atopic dermatitis can be a superimposed segmental manifestation of this polygenic disorder. Knowledge of this phenomenon, presence of less severe, disseminated, symmetrically distributed lesions of the same disorder and, if necessary, histology are helpful in the differential diagnosis from other linear inflammatory diseases.

KEYWORDS

atopic dermatitis, genetics, linear arrangement, superimposed segmental mosaicism of polygenic skin disorders

1 | INTRODUCTION

Common skin disorders with a polygenic background sometimes show a linear or otherwise segmental distribution of rather pronounced involvement that may either represent an isolated disorder or be superimposed on less severe, disseminated, symmetrically distributed lesions of the same disorder.¹ This last phenomenon is known as superimposed segmental manifestation. The idea that loss of heterozygosity may explain a segmental arrangement of a polygenic skin disorder was first presented in 1991, with linear psoriasis as a paradigm.² In the meantime, the number of disorders exemplifying this concept has considerably increased.¹ Case reports indicating superimposed segmental mosaicism have been found in dermatomyositis,³ psoriasis,⁴ lichen planopilaris,⁵ vitiligo⁶ and atopic dermatitis. Six reports of superimposed linear atopic dermatitis have been published thus far⁷⁻¹²; we describe one additional case.

2 | CASE REPORT

A 7-year-old girl presented with a segmental arrangement of eczematous lesions affecting the right arm since one year.

In the last 2 months, several nonsegmental eczematous lesions of similar appearance were noted on the anterior part of the neck, on the face and in the popliteal and cubital folds. During the observation period of 1 year, the lesion changed in severity but never disappeared completely, unless treated with a corticosteroid ointment. However, the dermatitis always recurred after cessation of therapy.

Family history revealed asthma in the maternal grandfather and allergic rhinoconjunctivitis in the father. No family members suffered from atopic dermatitis.

Physical examination showed erythematous macules, papulovesicular lesions, infiltrated plaques and excoriations, most pronounced in the face, the neck, the upper part of the trunk (Figure 1), and the right arm (Figure 2).

3 | DISCUSSION

Atopic dermatitis is a multifactorial chronic pruritic inflammatory skin disease, in which both genetic and environmental factors seem to be important in disease expression.¹³

The diagnosis of atopic dermatitis is clinical, based upon history, morphology and distribution of skin lesions (varying according to age), and associated clinical signs.



FIGURE 1 Diffuse eczematous lesions in face, neck, and upper part of the trunk



FIGURE 2 Eczematous lesions following a Blaschko linear pattern on the right forearm

As several other primarily symmetric skin diseases, atopic dermatitis may, on rare occasions, manifest itself more prominently along the Blaschko lines. This phenomenon is known as superimposed segmental manifestation, as a

counterpart of type 2 segmental mosaicism of monogenic skin diseases, and has tentatively been explained by the “ $n + 1$ ” rule.¹⁴ In a given case, the unknown number of predisposing heterozygous alleles would be n . Within the superimposed segmental involvement, their number would be $n + 1$ because either loss of the corresponding wild-type allele at one of the predisposing genes may have happened at an early developmental stage or a postzygotic new mutation would have occurred at an additional predisposing gene locus.

The concept of early loss of heterozygosity offers a plausible explanation as to why the segmental involvement tends to appear at a rather young age, often precedes the development of milder, nonsegmental lesions of the same disorder, and why the segmental lesions are notoriously difficult to treat. On the other hand, the theory of isolated versus superimposed segmental manifestation may help to elucidate the origin of polygenic skin disorders at the molecular level.¹

Superimposed linear atopic dermatitis may be more common than reported. This segmental distribution may pass unremarked, and cases of segmental dermatitis have been diagnosed with and published as other conditions, mainly lichen striatus¹⁵ and blaschkitis,¹⁶ especially when typical, symmetric AD lesions and/or extracutaneous signs of atopy are absent.

In conclusion, the linear manifestation most likely reflects the clonal outgrowth of a population of cells harboring a postzygotic mutation that increased the predisposition to atopic dermatitis. Knowledge of this phenomenon, presence of less severe, disseminated, symmetrically distributed lesions of the same disorder and, in case of doubt, histology are helpful in the differential diagnosis from other linear inflammatory diseases.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

JVG and RG: involved in patient management and in writing, editing, and reviewing of the manuscript.

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How to cite this article: Van Gysel J, Grimalt R. A linear lesion in a child with atopic dermatitis: Not a coincidence. *Clin Case Rep.* 2019;7:1667–1669. <https://doi.org/10.1002/ccr3.2322>