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Blaschkolinear Congenital Melanocytic Nevus

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Key Words

Congenital melanocytic nevus · Blaschko's lines · Mosaicism

Congenital nevi affect 1% of the newborns. We observed 1 linear case following Blaschko's lines.

A 14-year-old male presented with a congenital melanocytic nevus of the right leg present at birth and secondarily partially covered by terminal hair. The nevus was composed of one major plaque 15 cm in diameter associated with few smaller ones from 1 to 6 cm in diameter (fig. 1). The other leg was not affected. The nevus patches were arranged in a linear pattern following the lines of Blaschko on the posterior face of the right leg and thigh and covered 3% of the total body surface. The patient had other

common acquired nevi and an otherwise normal clinical examination.

Congenital melanocytic nevi are often classified according to their size [1]. Another attempt of classification in two groups has recently been proposed: type I, the most frequent, with a single lesion involving one anatomic area and type II, less common, with a larger lesion and dozens of smaller ones, with a similar pattern on histological analyses, involving more than one anatomic area [2].

The lines of Blaschko were described in 1901 by Alfred Blaschko [3]. They are thought to follow the dorsum-to-ventral migration of the epidermal cell clones from the neural crest to the genetically programmed definitive skin location [4]. Several observations of common monogenic or polygenic skin disorders following Blaschko's lines have been reported [4, 5]. Linear congenital melanocytic nevus as seen in our case has been reported once [5].

Focusing on large congenital melanocytic nevi, Effendy and Happle [6] reported in 1992 the first case of congenital melanocytic nevus, constituted of an association of multiple nevi with linear arrangement involving an arm, following the lines of Blaschko. This case and the present patient could be defined as having type II congenital melanocytic nevi [2]. More recently, a study has analyzed 92 cases of congenital melanocytic nevi and has claimed that 39 could be considered to match the lines of

Fig. 1. Congenital melanocytic nevus of the right leg arranged in a linear pattern following the lines of Blaschko in a 14-year-old male.

Color version available online

Blaschko [7]. This is the first study to systematically investigate the skin distribution of these nevi according to Blaschko's lines in congenital nevi, excluding the giant subtype and aggregate of speckles. But this study seems debatable as the sizes of the lesions were not fully reported and the pictures shown did not present evidence of Blaschko's distribution. Furthermore, it seems difficult to prove that small lesions truly match this pattern [7].

A recent case of acquired numerous melanocytic nevi arranged in a linear pattern on the left arm has been described and considered as an unusual type of mosaicism of melanocytic disorders [8]. The concept of mosaicism could give an attractive explanation of the linear distribution of the nevus as observed in our case and those of Happle and Effendy [6]. Indeed, a clonal outgrowth of cells, resulting from a mutation occurring at an early stage of embryogenesis, seems to be the support of the clinical features of these nevi following the lines of Blaschko. This hypothesis is not yet supported by a molecular study. Nonetheless, even though demonstrated in a keratinocytic model, a recent study identified a putative molecular basis of linear nonepidermolytic, nonorganoid keratinocytic epidermal nevi [8]. In this study, linear epidermal nevi could be linked to postzygotic mutations in the fibroblast growth factor receptor 3 gene (tyrosine kinase receptor), which occurred in a single keratinocyte stem cell at an early stage of the embryonic development [9]. Although the molecular signature of congenital melanocytic nevi remains unclear, the clinical features of the linear nevus described herein support the hypothesis of such a postzygotic genetic incident of an unknown gene causing the proliferation of a clone of melanocytic cells with a nevus' phenotype at an early stage of embryogenesis.

We report herein one additional case of nevus following Blaschko's lines. This particular distribution could result from a possible postzygotic mosaicism among Blaschko's lines, although molecular evidence is still lacking in this setting and needs to be investigated.

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