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Poïkiloderma with Neutropenia (Clericuzio Syndrome) in a Brother and Sister

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ABSTRACT

Congenital poïkiloderma is infrequent and immunity deficiency associated with poikiloderma is poorly reported. The association of congenital poïkiloderma and neutropenia known as Clericuzio syndrome is rare. The family involvement of this entity is exceptional. Analyzing two clinical cases in a brother and sister, the authors underline the fact that the cutaneous manifestations made possible to specify the etiology of this familial congenital neutropenia after several years of diagnostic wandering.

Keywords: Poïkiloderma, Neutropenia, Clericuzio, Syndrome, Black skin

INTRODUCTION

Congenital poïkiloderma is an infrequent dermatosis. The clinical profile is very variable and the diagnosis is not always easy. It may associate with mucous, phanerian and ocular involvement or integrate into complex syndromes [1]. Immunity deficiency associated with poikiloderma is poorly reported. The association of congenital poikiloderma and neutropenia known as Clericuzio syndrome is rare [2]. It is a genodermatosis with autosomal recessive transmission. The family involvements of this nosological entity are exceptional. We report an observation in a brother and sister.

OBSERVATION

First Clinical Case

MK, 4 years old was born from non-consanguineous parents. She had consulted at the age of 6 months in the department for a pruritic dermatosis. She had recurrent cutaneous abscesses since the age of two months, associated with pleuropulmonary and gastrointestinal infections. The cutaneous-mucous examination found eczematous lesions of the limbs and trunk. The biological balance showed hypochromic microcytic anemia, neutropenia at 250 cells / mm3. The patient had been treated with topic steroïd and oral antibiotic therapy. She then did not return to the appointments of controls for more than 3 years. In font of recurrent infections, she reportedly had numerous consultations in pediatrics where the various biological assessments carried out revealed a permanent neutropenia. She was then treated with granulocyte growth factors. At 4 years old, we saw her again at the dermatological consultation. The clinical examination finds growth retardation associated with a diffuse poïkilodermia. Indeed she had in most of the part of her body an hyperpigmentation associated whith an hypopigmentation and a skin atrophy (**Figure 1).** She had no palmo-plantar keratoderma but a photosensitivity. The diagnosis of Clericuzio syndrome was retained. There were no teeth abnormalities. The hairs and the mucous membranes were normal. A topical steroid for the eczematous lesions and a moisturizing cream were prescribed. The hematological disorder was follow up by an hemato-pediatrician. The treatment leads to a stabilization of the poikiloderma and the decreasing of the infections.

Second Clinical Case

MK's elder brother, 7 years old, was born at the end of a monofetal pregnancy. He presented from the age of 2 months digestive disorders such as gastroenteritis and repeated eczematous lesions. He had also been hospitalized several times for multiple infections including bacterial pneumonia, urinary tract infections and cutaneous abscesses.

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The various biological examinations performed revealed a permanent neutropenia ranging from 300 to 500 elements / mm3. At 6 months the child presented a progressive installation of poïkiloderma associated with plantar hypopigmentation macules resembling Poikilodermie-Neutopenie syndrome of his younger sister (**Figure 2).** A photosensitivity was noticed but he had no palmo-plantar keratoderma. The hairs and mucous membranes were also unremarkable. He had received the same treatment as his sister with a stabilization of the lesions. No other relatives have similar symptoms. Multidisciplinary management and follow up was recommended with parental education in photoprotection, regular dermatological and hematological monitoring.

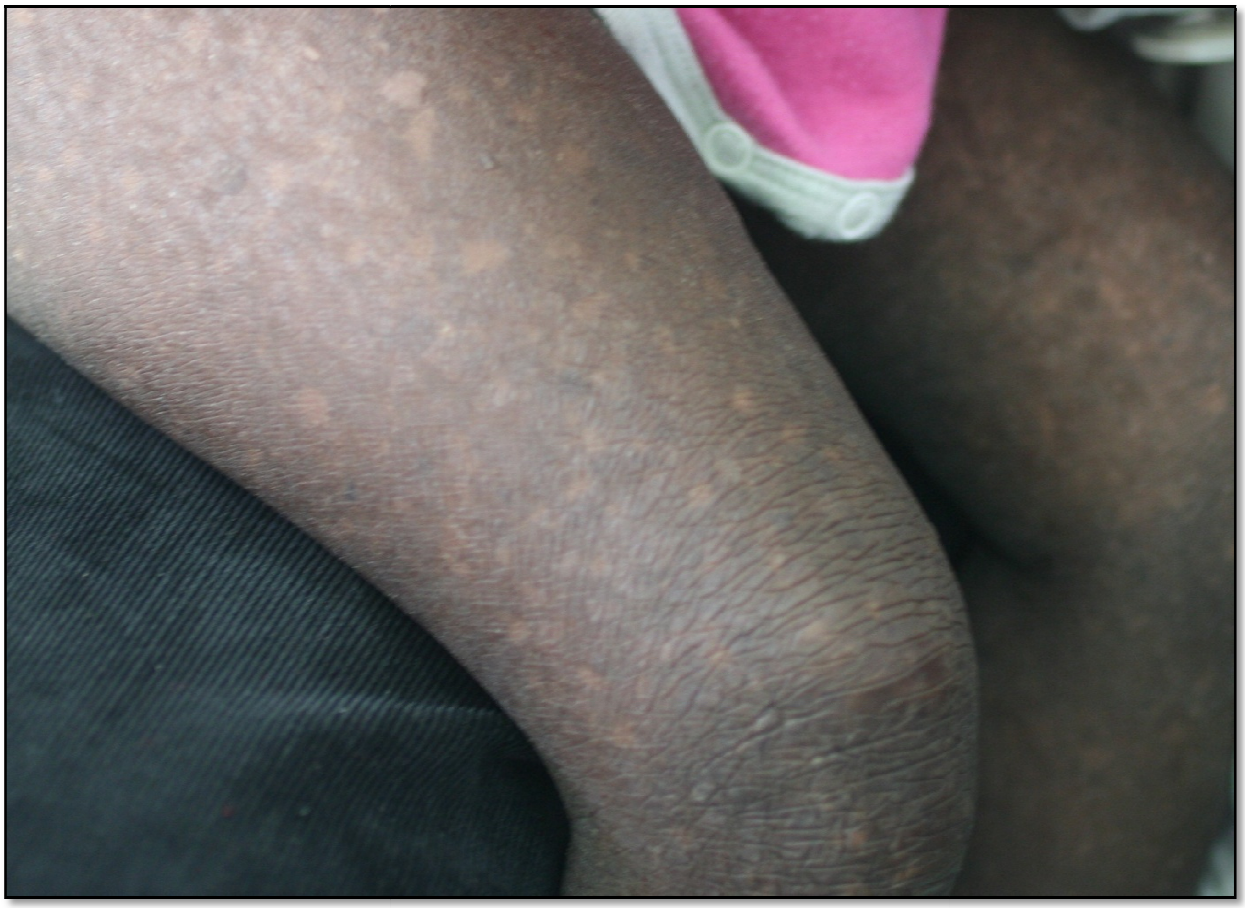

Figure 1. Case 1: Poïkilodermia of the limbs of the sister

DISCUSSION

Congenital poïkiloderma is rare. It may be the sign of some genodermatosis such as Rothmund-Thomson syndrome, Bloom syndrome, xerodermapigmentosum and congenital dyskeratosis [3]. Poikiloderma-Neutropenia syndrome was described for the first time in 1991 among the peoples from Navajo by Clericuzio. This syndrome is characterized by an eczematous onset of the skin, and secondarily by a poïkilodermal aspect of the skin, which predominates at the extremities with centripetal evolution [4]. It is frequently associated with recurrent bacterial and viral infections and hematological damage characterized by permanent neutropenia, leucopenia and thrombocytopenia. Patients often show a small size, pachyonychia and palmoplantar hyperkeratosis [5,6]. Bone abnormalities as well as malignant transformations have been reported [7]. This dominant autosomal genodermatosis is characterized by a mutation of the C16ORF 57 gene [8,9]. Management is difficult and consists essentially in the prevention of infections. A better understanding of genetic abnormalities will certainly lead to the development of targeted therapies. In this singular observation, the cutaneous manifestations made it possible to specify the etiology of this congenital neutropenia in a brother and sister after several years of diagnostic wandering..

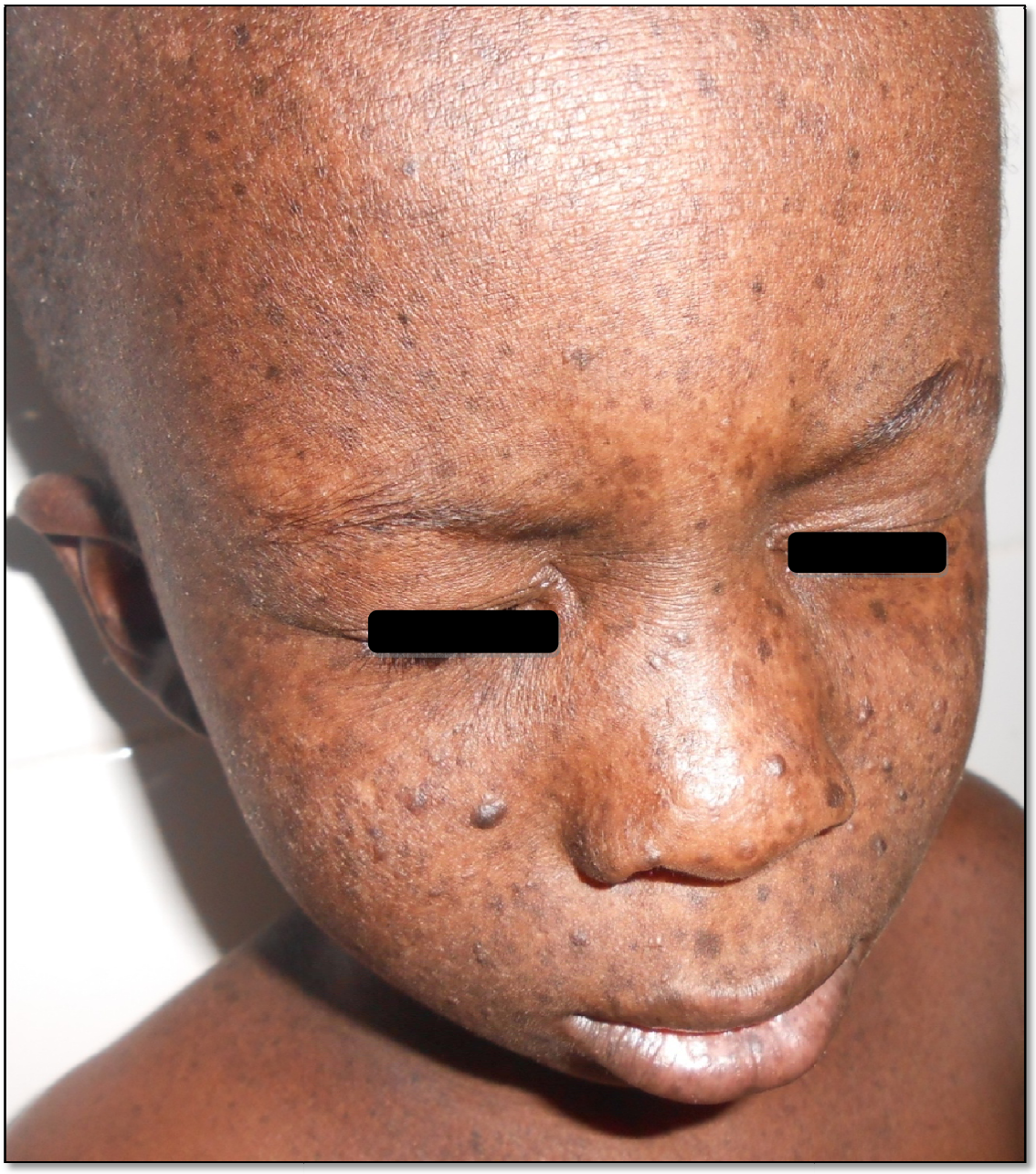

Figure 2. Case 2: Poïkilodermia of the face of the elder brother

CONCLUSION

Poikilodema-Neutropenia syndrome is a severe and rare genodermatosis. This reported case shows that it is ubiquitous. In front of a congenital poïkilodermal condition, the search of neutropenia and other signs of Clericuzio syndrome is necessary in order to lead the patient early to a multidisciplinary care.

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