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Case Report

Multiple Nail Candidosis in a Child with APECED Syndrome

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Abstract

The APECED syndrome (autoimmune polyendocrinopathy, candidosis, ectodermal dystrophy) is a rare autosomal recessive disorder that develops in early childhood and results in tissue-specific multiorgan autoimmunity, leading to the hypofunction of multiple glands. Is clinically defined as the presence of at least two components of the classic triad of hypoparathyroidism, adrenal insufficiency, and mucocutaneous candidiasis. We report a case in a child, illustrating the importance of dermatological disorders.

Keywords: APECED syndrome; ,Mucocutraneous candidosis; AIRE gene

Introduction

The APECED syndrome (autoimmune polyendocrinopathy, candidosis, ectodermal dystrophy) is a rare condition of autosomal recessive inheritance linked to mutations of the AIRE gene [1;2]. We report a case in a child, illustrating the importance of dermatological disorders.

Case Report

A 9-year-old boy, from a second-degree consanguineous marriage, followed in pediatrics for APECED syndrome confirmed by the presence of the mutation of the AIRE gene, with autoimmune hypoparathyroidism, nephrocalcinosis, autoimmune hepatitis and nail dystrophies evolving since three years.

Dermatologic examination revealed pachyonychia with xantonychia, leukonychia, distolateral onycholysis and hyperkeratosis under the nail of the twenty nails. The mycological specimen returned positively to candida albicans. Oesogastroduodenal fibroscopy returned normal. Systemic antimycotic treatment has been proposed.

Discussion

The APECED syndrome is a rare, serious genetic disease of autosomal recessive inheritance, which must be considered in the face of recurrent cutaneous candidosis and mucosal candidois infections with a characteristic triad consisting of recurrent mucocutaneous candidosis, hypoparathyroidism and autoimmune adrenal insufficiency [3;4]. The care is difficult, multidisciplinary based on patient education, hormonal supplementation and long-term systemic antimycotic therapy [5].

Conclusion

Knowing the cutaneous disorders of the APECED syndrome allows the dermatologist to diagnose it early, before the appearance of the serious endocrine disorders.

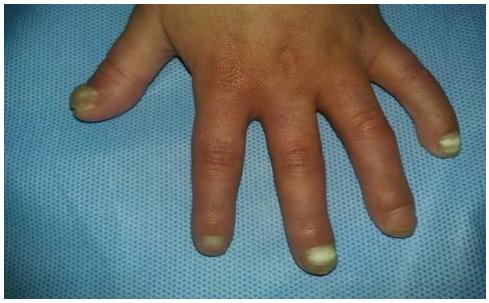

Figure 1: Nail candidosis of the left hand

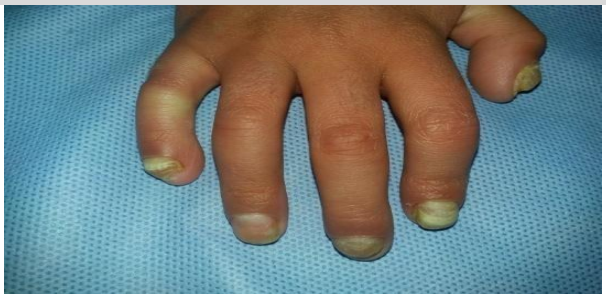

Figure 2: Nail candidois of the right hand

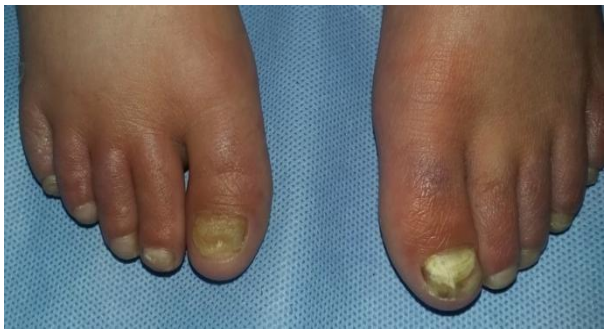

Figure 3: Candidosis of the feet

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