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ORIGINAL ARTICLE

Acrodermatitis enteropathica or malabsorption syndrome secondary to cystic fibrosis?

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Zinc Deficiency.

Abstract

Cystic fibrosis (CF) is a multi-systemic disease, which can affect many organs, such as the gastrointestinal system, making individuals with CF have difficulty absorbing many nutrients, leading to malabsorption syndrome. Another disease whose repercussions derives from bowel malabsorption is the acrodermatitis enteropathica, which is a rare genetic autosomal recessive disorder of zinc deficiency. We present a case of a child born at term, low birthweight, without clinical complications at birth. He was referred to the CF center at 5 months of age, due to alteration with the newborn screening for CF and the sweat test, failure to thrive and clinic steatorrhea. Based on the diagnostic hypothesis of acrodermatitis enteropathica, we initiated oral supplementation with zinc gluconate 1mg/kg/day, empirically, for serum level was unavailable. Therapeutic measures for CF were introduced, such as: hyper caloric diet, vitamin supplements, pancreatic enzymes, increase in salt and water input. Rapid and meaningful improvement of the skin lesions. The case shows the importance to consider acrodermatitis enteropathica and CF as differential diagnosis.

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INTRODUCTION

Cystic fibrosis (CF) is an autosomal recessive genetic disease characterized by dysfunction of the cystic fibrosis transmembrane conductance regulator (CFTR) gene, which encodes a chloride transmembrane conductance regulatory protein. In Brazil, it is estimated that the incidence of cystic fibrosis is 1:7,576 live births. It is a multisystem disease that can affect several organs of the body, including the gastrointestinal system. Due to the change in chloride channels, individuals with CF have difficulty absorbing various nutrients, which can lead to malabsorption syndrome[1]. Acrodermatitis enteropathica is a genetic, autosomal recessive disease that leads to a deficiency in intestinal zinc absorption[2]. Zinc is one of the main chemical elements in the body and can exert catalytic, structural and regulatory functions[3,4].

CASE REPORT

Male child, born at term, low weight, without clinical complications at birth, with altered immunoreactive trypsin dosage in the neonatal screening test . At 2 months of age , bullous skin lesions appear, with scaling and crusts. Evolving with difficulty in weight and height gain and clinical steatorrhea. Only at 4 months, sweat chlorine was measured by the conductivity method, with an abnormal result. Due to this change, the child was referred to a pediatric cystic fibrosis (CF) referral center at 5 months of age. On physical examination: weight 4770g (z -4.36), length 58.4 cm (z -4.11) and body mass index of 14 kg/m² (z -2.65), thinning hair. In the occipital region, there was an erythematous lesion associated with hematic crusts. In the perineal region, exulcerated and scaly lesions and, in the lower limbs, xerotic, rough skin and also areas of desquamation, as shown in figure 1. With the diagnostic hypothesis of acrodermatitis enteropathica, oral supplementation with zinc gluconate 1mg/kg/day was started, empirically, because the serum level was unavailable. Other therapeutic measures for cystic fibrosis were initiated, such as: hypercaloric diet, vitamin supplementation, pancreatic enzymes and increased intake of salt and water in the diet. Fast and significant improvement of the lesions was observed after about a week of use. Zinc replacement was discontinued after 55 days, and the patient did not present new skin lesions, remaining stable. The hypothesis of CF was confirmed after the measurement of chlorine in sweat by the coulometry method.

COMMENTS

Cystic fibrosis (CF) may present with exocrine pancreatic insufficiency and should be suspected in the presence of steatorrhea, chronic diarrhea, low weight gain and signs of hypovitaminosis[1]. Acrodermatitis enteropathica (AE) is a genetic disease, with autosomal recessive inheritance, which leads to a deficiency in intestinal zinc absorption. Clinical manifestations are characterized by eczematous and erosive dermatitis, alopecia, difficulty in gaining weight, growth retardation, anemia, greater predisposition to infections, diarrhea, among others. The treatment of choice is supplementation of zinc, in an average dose of 1 to 2 mg of elemental zinc/kg/day[2-4]. The patient in question had exulcerated and scaly skin lesions, with distribution in the extremities and perineum, as seen in cases of AE. It is known that in the absence of treatment for this dermatitis with zinc deficiency, patients develop alopecia and diarrhea[2], as in the case mentioned above. The signs and symptoms presented in this case could correspond to acrodermatitis enteropathica, however, in the presence of the diagnosis of CF, these manifestations were related to the malabsorption syndrome secondary to the underlying disease, so much so that, after the suspension of zinc replacement, there was no there was a return of the lesions as would be expected in cases of acrodermatitis enteropathica. The case described highlights the importance of considering acrodermatitis enteropathica and CF as differential diagnoses. It is believed that zinc supplementation contributed to faster clinical improvement of the patient, in addition to other therapeutic measures of CF.

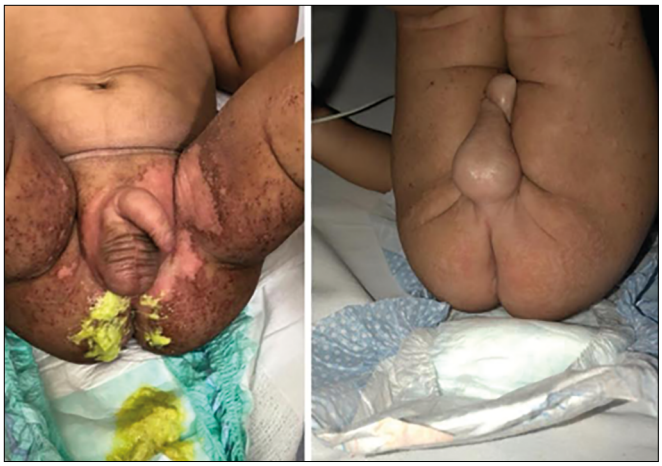

Figure 1. A. at the first appointment at the Cystic Fibrosis Reference Center of the Federal District, with skin lesions and steatorrhea; **B.** 30 days after initiation of CF treatment and zinc replacement.

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