Acquired acrodermatitis enteropathica from a ketogenic diet

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INTRODUCTION

Acrodermatitis enteropathica (AE) is a condition of zinc deficiency and is either congenital or acquired. The congenital form is caused by an autosomal recessive genetic defect resulting in decreased zinc absorption, whereas acquired AE is due to reduced zinc intake or absorption from other causes. Up to one-third of some populations in developing countries are at risk of zinc deficiency, which has been associated with increased disease and stunting of growth in children.[1] In developed countries, AE most commonly affects individuals with risk factors such as alcoholism, vegetarianism, premature infancy, and malnourishment.[1] Here, we present a unique case of AE in a child on a ketogenic diet.

CASE DESCRIPTION

A 11-year-old boy with a history of hypoxic ischemic encephalopathy and severe epilepsy since infancy presented to the dermatology clinic with perioral, periorbital (Fig 1), perigenital (Fig 2), and acral (Fig 3) eruption for the past 2 months. His mother had also been noticing mild hair loss and more foul-smelling loose stools. Medications included levetiracetam and clobazam. The patient had been adhering to a ketogenic diet for 3 months, which, according to his mother, resulted in fewer seizures. Due to dysphagia and epilepsy, the patient was administered a liquid ketogenic formulation consisting of a complete amino acid mixture and both long-chain and medium-chain triglyceride emulsions through a gastrostomy tube. Mineral and vitamin supplementations were recommended but not given. Laboratory studies showed zinc levels of 15 mcg/dL (normal levels, 50.0−120.0 mcg/dL). A biopsy demonstrated superficial epidermal necrosis with cytoplasmic vacuolization and scattered dyskeratotic keratinocytes (Fig 4) consistent with the clinical impression of acquired zinc deficiency. The family discontinued the ketogenic diet and initiated treatment with zinc gluconate 50 mg/day and topical emollients. His symptoms began improving within 3 days and had completely resolved one month later.

Abbreviation used:

AE: Acrodermatitis enteropathica

DISCUSSION

Acquired AE has been associated with many causes of malnutrition, including veganism, alcoholism, anorexia nervosa, and bariatric surgery; in our search of the literature we could not find a ketogenic diet as a specific cause. The classic ketogenic diet is a high-fat, low-carbohydrate diet, comprising primarily long-chain triglycerides, with a 3:1-4:1 ratio of grams of fat to carbohydrates and protein.[2] This diet has been used to treat epilepsy since the early 1920's, and although its popularity initially dwindled upon the advent of anti-epileptic medications in the late 1930's, it later became an established therapy for medication-resistant epilepsy.[3] The ketogenic diet has also more recently evolved into a worldwide health trend due to its proven ability to cause quick and substantial weight loss,[4] but not without consequence. Studies have shown that those adhering to a strict ketogenic diet should have routine monitoring of micronutrients with appropriate supplementation, since the diet can

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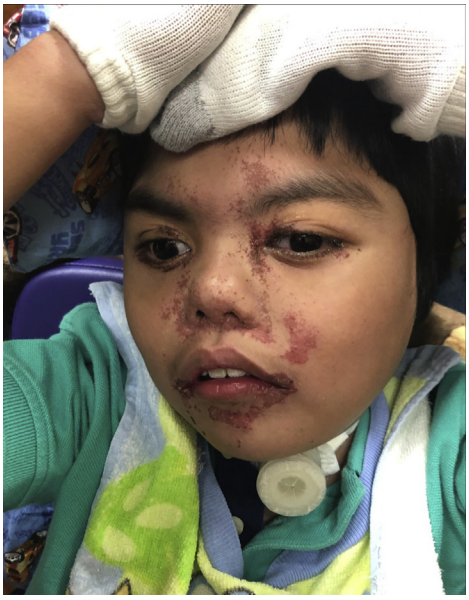

Fig 1. Perioral and periorbital dermatitis.

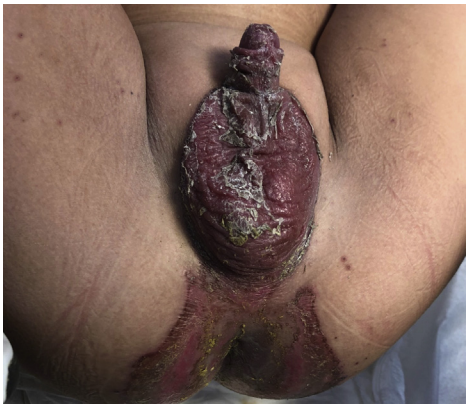

Fig 2. Perigenital dermatitis extending from the penis and scrotum to the perianal region.

lead to various nutritional deficiencies, including zinc and ascorbic acid.[5,6] The diet has also been reported to cause prurigo pigmentosa.[7,8] The liquid form of the ketogenic diet has proven to be safe and effective for pediatric patients with a gastrostomy tube[9]; however, nutritionally complete formulations supplemented with nutrients and vitamins (including zinc) should be used.

Zinc plays multiple important physiologic roles; it is a catalyst for many metabolic reactions, a structural component of numerous proteins, and is involved in gene expression and other regulatory functions.[1] Zinc deficiency, whether acquired or congenital, can lead to the typical signs and symptoms of AE. AE classically presents with a triad of alopecia, dermatitis, and diarrhea. However, only 20% of the cases present with all 3, as observed in the present case.[10] More advanced presentations include growth delay, hypogonadism, hypoguesia, poor wound healing, and increased risk for secondary bacterial infection of skin lesions.[1] Diagnosis usually relies on a thorough patient history, physical exam, and investigation of serum zinc levels, but a high index of suspicion is necessary. Biopsy may be helpful, but histopathology is identical to other forms of nutritional deficiency dermatitis, such as niacin deficiency and necrolytic migratory erythema.[1] Practitioners should be aware of the need for vitamin and mineral supplementation in patients on a ketogenic diet, especially those on a liquid formulation. Additionally, particularly given the rising popularity of the ketogenic diet, it is important to be aware of its potential side effects and their dermatologic manifestations; these include the possibility of prurigo, scurvy, and, as in our patient, acquired AE.

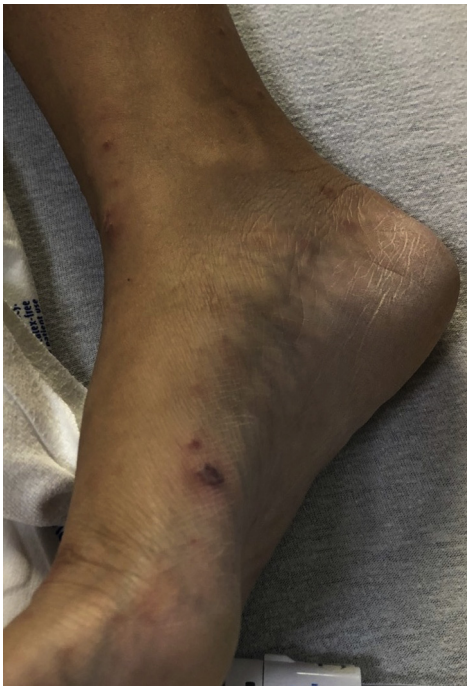

Fig 3. Acral dermatitis.

Conflicts of interest

None declared.

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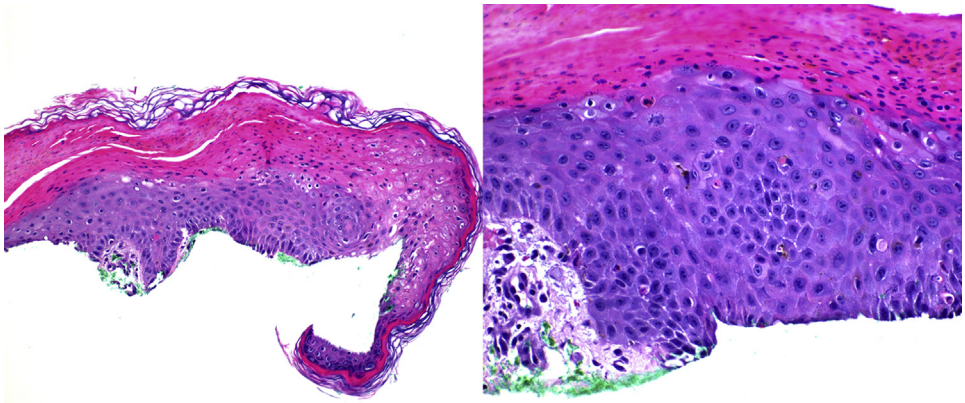

Fig 4. Hematoxylin and Eosin stain demonstrated superficial epidermal necrosis with cytoplasmic vacuolization and scattered dyskeratotic keratinocytes consistent with the clinical impression of acquired zinc deficiency.

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