Acquired Acrodermatitis Enteropathica Syndrome in a Kidney Transplant Recipient: A Case Report

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ABSTRACT

Acrodermatitis enteropathica syndrome (AE) is a clinical entity that results in severe zinc deficiency. It can be genetic or acquired. Acquired AE has been reported in patients with chronic liver disease, malabsorption syndrome, sickle cell anemia, and chronic renal failure. We present a kidney transplant recipient with skin rash and watery diarrhea. The patient had low serum zinc levels, which quickly resolved after zinc supplementation. Skin biopsy showed cytoplasmic pallor and vacuolization and ballooning degeneration of keratinocytes within the superficial epidermis, which may have led to confluent necrosis of keratinocytes. Large amounts of keratinosome-derived lamellae were found in the intercellular spaces in the keratinized area, probably related to disturbance of keratinosome metabolism due to zinc deficiency.

ACRODERMATITIS enteropathica syndrome (AE) is a clinical entity that results in severe zinc deficiency. It can be genetic or acquired. The inherited form usually develops early in life and is characterized by periorificial dermatitis, alopecia, and diarrhea. Other causes of zinc deficiency are called acquired AE, which has been reported in patients with chronic liver disease, malabsorption syndrome, sickle cell anemia, and chronic renal failure [1]. We present a kidney transplant recipient with acquired AE, which resolved after zinc supplementation.

CASE REPORT

A 21-year-old man was admitted to clinic with skin rash and watery diarrhea. He had received a deceased donor kidney transplant 2 years ago because of end-stage renal failure secondary to nephronophthisis. His immunosuppression regimen was mycophenolate mofetil 1 g twice daily, tacrolimus 1 mg twice daily, prednisolone 5 mg once daily. On physical examination, eczematous pink scaly plaques, psoriasiform, and pustular, vesiculobullous lesions were noticed on hands, feet, and periorificial areas (Fig 1).

Laboratory investigations disclosed normal levels of glucose, uric acid, cholesterol, triglycerides, aspartate amino transaminase, alanine amino transaminase, alkaline phosphatase (ALP), and bilirubin. Serum creatinine, sodium, potassium, calcium, and phosphorus were in normal range. Hemoglobin was 11 g/dL, and white blood cell and platelets were also normal.

We suspected psoriasis, and a local treatment with fluticasone propionate was started after skin biopsy. Histopathologic examination of left foot biopsy revealed a spongiotic dermatitis characterized by intraepidermal vacuolization (Fig 2), showing thickening in the keratin layer with focal parakeratosis and intraepidermal vesicle formation with neutrophils and eosinophils in acanthotic epidermis. Due to the distribution and unusual appearance of the lesions, AE was suspected, and we measured the serum zinc levels, which were 0.23 μg/mL (normal value: 0.92 to 1.44 μg/mL). We diagnosed AE.

Zinc therapy was initiated (400 mg daily of elemental zinc), with rapidly favorable evolution: the lesions and diarrhea disappeared after 5 to 6 days of treatment. Two years after completion of therapy, the patient remains asymptomatic with no recurrence of skin lesions (Fig 3).

DISCUSSION

AE was first described in 1936 by Brandt as a skin lesion around the trunk, extremities, and natural openings of the body, associated with diarrhea, alopecia, atrophy of the nail, and photophobia. AE is a disorder associated with poor zinc

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Fig 1. Eczematous pink scaly plaques, psoriasiform, and pustular, vesiculobullous lesions on hands and feet.

absorption and is classified as both genetic and acquired forms. The genetic form is a rare autosomal recessive condition and arises in infancy or early childhood. The mechanism of AE is considered to be mutations in the *SLC39A4* gene on chromosome 8q24.3, namely, the gene encoding zinc-transporting protein [2,3]. In our case, we diagnosed our patient as an acquired AE because of unremarkable family history and late symptoms. Acquired AE may present at any age from infancy through adulthood and may occur due to dietary disorder including alcoholism and gastrointestinal disease including pancreatic insufficiency, inflammatory bowel disease, malabsorption, burns, malignancy, chemotherapy, and pregnancy.

Acquired AE has previously been reported in patients with end-stage renal failure on maintenance dialysis; patients had low concentrations of zinc in plasma, leukocytes, and hair. The disorder usually remained asymptomatic; however, uremic hypogeusia and gonadal hypofunction resolved after zinc supplementation [4]. To the best of our knowledge AE has not been described in a renal transplant recipient before. In

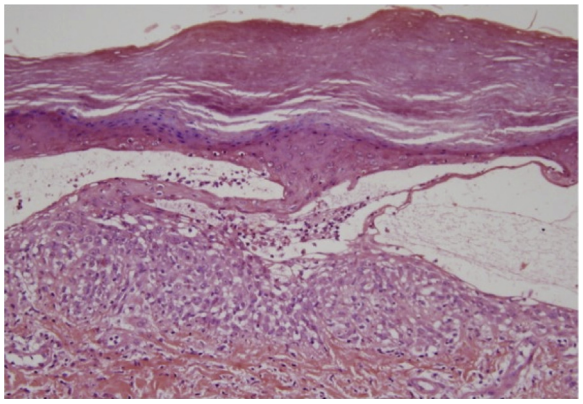


Fig 2. Histopathologic examination of left foot biopsy revealed spongiotic dermatitis characterized by intraepidermal vacuolization.

our case, poor dietary intake or tubular zinc wasting in the early postoperative period caused zinc deficiency.

ALP activity in patients with AE is generally low, although normal and even elevated levels have been reported [5]. In our patient, serum ALP was found to be normal at the time of diagnosis (77 U/L).

Skin biopsy showed cytoplasmic pallor, with vacuolization and ballooning degeneration of keratinocytes within the superficial epidermis, which may subsequently lead to confluent necrosis of keratinocytes. Large amounts of keratinosome-derived lamellae were found in the intercellular spaces in the keratinized area, probably related to disturbance of keratinosome metabolism due to zinc deficiency [6].

In summary, we present a kidney transplant recipient with acquired AE, which quickly resolved after zinc supplementation. AE should be considered in renal transplant



Fig 3. Lesions disappeared after treatment.

recipients who present with skin rash and diarrhea. AE and zinc deficiency can be diagnosed by distribution and appearance of the lesions, measuring plasma or serum zinc levels, histopathologic examination, and revealing symptoms by administering a zinc load.

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