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Acrodermatitis enteropathica in a pair of twins

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Abstract

Background: Acrodermatitis enteropathica (AE) is a rare autosomal recessive metabolic disorder. First described by Brandt in 1936 and was named by Danbolt. A mutation in the SLC39A4 gene on chromosome 8 q24.3 is responsible for this disorder, which encodes zinc transporter Zip4. The diagnosis is made by the clinical presentation and histopathology and laboratory tests. In this case, we reported a twin presented with a typical rash and low zinc level. To our knowledge, very few cases reported as a twin with typical acrodermatitis enteropathica presentation.

Main observations: Four months old twins both females, first children of a non-consanguineous marriage. The twins were born at term, caesarian section, with no complications. Presented with erythema, scaling, crusting and oozing over perioral, perianal areas, hands and feet of 2-3 week duration. The lesions started around the same time for both children with a history of intermittent diarrhea, and hair loss. There were no nail changes or neurological deficit or myopathy. There was a history of recent weaning from breast milk and now both children on formula feeds, ragi, fruits. There was no other significant history of other medical problems in the patients or in their family. On examination, erythema, scaling, crusting and oozing over perioral, perianal areas, hands and feet was seen. Minimal diffuse alopecia was noted. Nails were normal. No other abnormalities were observed.
Clinical diagnosis of acrodermatitis enteropathica was considered and confirmed by low zinc levels (repeated plasma zinc levels were below 0.6 mcg/ml). The twins were managed with zinc supplementation 1 mg/kg/day. A significant improvement was seen within two weeks.

Conclusions: Early diagnosis of acrodermatitis enteropathica is essential for preventing complications. We report a rare case of typical clinical presentation of the disease developing simultaneously in twins. (*J Dermatol Case Rep*. 2016; 10(4): 65-67)

Introduction

Acrodermatitis enteropathica (AE) is an autosomal recessive metabolic disorder. First described by Brandt[1] in 1936. A mutation in the SLC39A4 gene on chromosome 8 q24.3 is responsible for the disorder.[2] The SLC39A4 gene encodes a transmembrane protein, which works as a zinc uptake protein. Acrodermatitis enteropathica is characterized by inflamed patches of dry red skin, which then become crusted and blistered before revealing a pustulent-eroded lesion. Typically this rash starts near the body's orifices before migrating to other sites. In this case, we reported a twin presented with a typical rash and low zinc level. To our knowledge, very few cases reported as a twin with typical acrodermatitis enteropathica presentation.

Case Report

Four months old twins both females, first children of a non-consanguineous marriage. The twins were born at term, caesarian section, with no complications. Presented with erythema, scaling, crusting and oozing over perioral, perianal areas, hands and feet of 2-3 week duration (Fig. 1-5). The lesions

started around the same time for both children with a history of intermittent diarrhea, and hair loss. There were no nail changes or neurological deficit or myopathy. There was a history of recent weaning from breast milk and now both children on formula feeds, ragi, fruits. There was no other significant history of other medical problems in the patients or in their family. On examination, erythema, scaling, crusting and oozing over perioral, perianal areas, hands and feet was seen. Minimal diffuse alopecia was noted. Nails and other mucosae were normal. Systemic examination was normal.

Scraping from the lesions was negative for candida and gram stain also did not show any significant organisms. Routine blood and urine investigation were within normal limits. VDRL and HIV ELISA were negative. Repeated plasma zinc levels were low (<0.6 mcg/ml) but alkaline phosphatase levels were normal. Clinical diagnosis of acrodermatitis enteropathica was considered and confirmed by low zinc levels. Skin biopsy was not taken because the diagnosis was clear clinically and because of the age of the patients. The twins were managed with zinc supplementation 1 mg/kg/day. An improvement was seen within about two weeks.

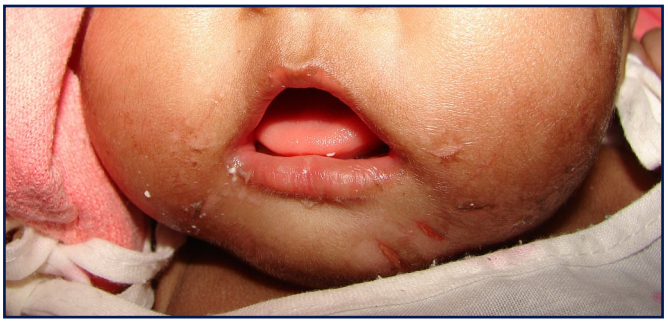

Figure 1
Erythema, scaling, crusting and oozing over perioral area.

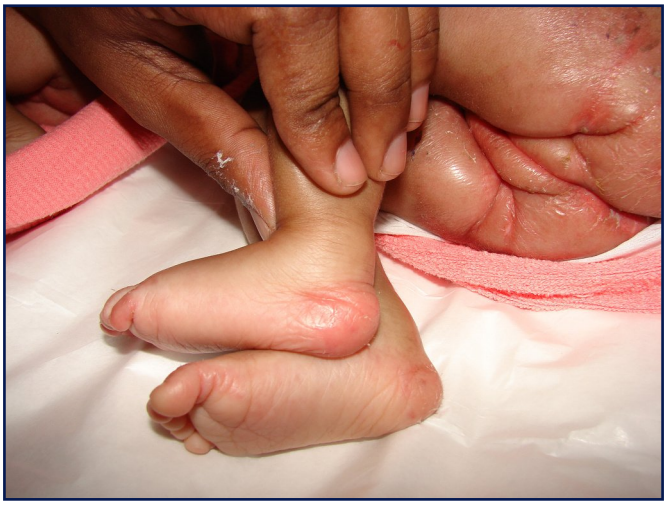

Figure 2
Erythema, scaling, crusting and oozing over feet and perianal areas.

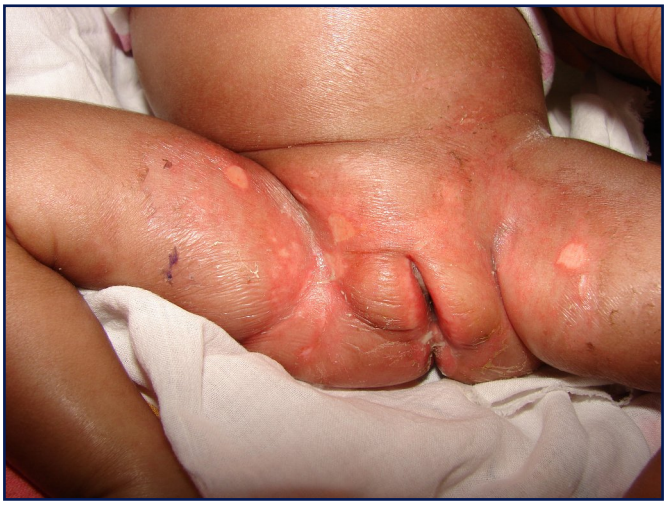

Figure 3
Erythema, scaling, crusting and oozing over perianal area.

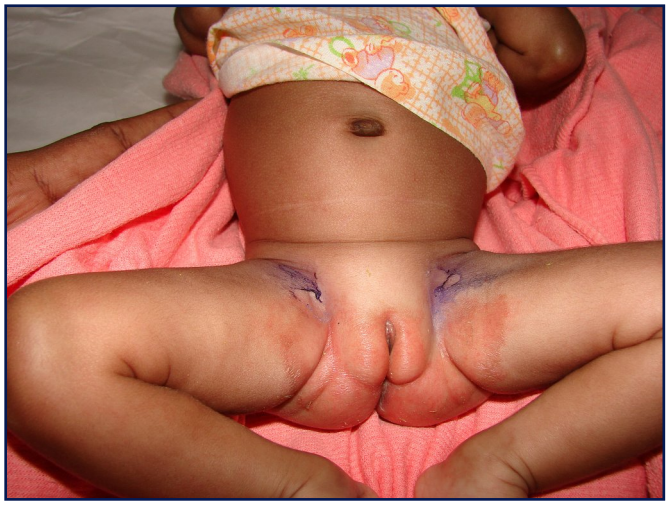

Figure 4
Erythema, scaling, crusting and oozing over perianal.

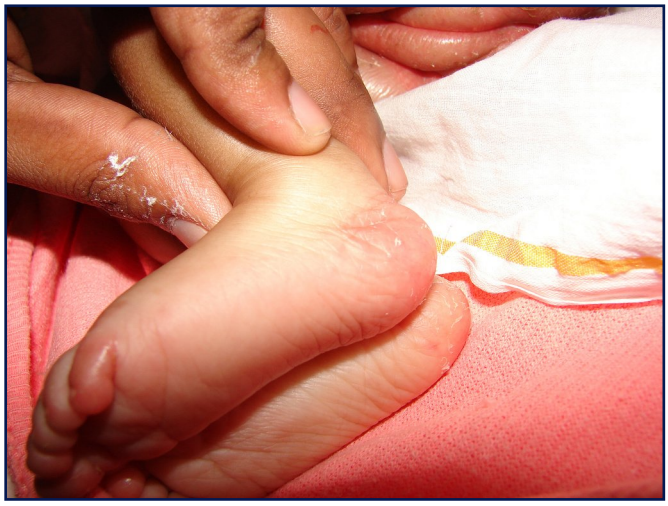

Figure 5
Erythema, scaling, crusting and oozing over feet.

Discussion

The optimal growth and development of infants require various micronutrients. Zinc is an essential trace element necessary for the proper functioning of all cells. It has a significant role in the metabolism of protein, carbohydrate, and vitamins.[3] The functions of zinc are divided into three categories: catalytic, structural, and regulatory.[4] Zinc deficiency is either hereditary or acquired. The hereditary deficiency is a condition called acrodermatitis enteropathica (AE). AE is an autosomal recessive mutation of SLC39A4 (solute carrier family 39 member 4) gene on chromosome 8q24.3; that determines a congenital partial or total deficiency of the zinc transporter protein zinc-ligand binding protein 4 (ZIP 4).[2] The clinical triad of AE is periorificial dermatitis, alopecia, and diarrhea. This triad is found complete in only 20% of reported cases.[5] In our case, the complete triad is present in both twin; however, other reported cases have an atypical presentation.[6,7] Diagnosis of AE is based on clinical symptoms and is confirmed by low plasma zinc levels and rapid clinical response to zinc supplementation. In other hand, many cases have normal zinc level with manifestations of AE. In our patients, zinc level was low for both cases, and this is similar to various cases reported.[6,8,9] However, normal zinc level was found in other cases.[10] In our case, AE was presented in a twin at the same time for both children and this probably reflects an underlying genetic predisposition for development of AE.

The limitations of our case report are that skin biopsy was not performed because of the age of patients. Also the patients did not do follow up after that. We present this case because of the interesting simultaneous occurrence in a pair of twins.

Conclusion

Acrodermatitis enteropathica is a rare disease; quick and early identification of skin manifestation are necessary for preventing complications. There are various presentations of this disorder. The simultaneous occurrence in twins possibly highlights the underlying genetic component in our case.

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