Acrodermatitis Enterohepatica: A Series of 5 Cases Seen In Aminu Kano Teaching Hospital Kano,nigeria.

Yahya AM, Yusuf SM

Division of Dermatology, Aminu Kano Teaching Hospital, Kano, Nigeria.

Corresponding Author: *Aishatu M. Yahya.* ***E-Mail:*** *aishayahya35@yahoo.com,*

Shehu M Yusuf. ***E-Mail:*** *shehumy@yahoo.com*

ABSTRACT

Acrodermatitis enterohepathica (ADE) is a rare form of zinc deficiency, characterized by perioral and acral dermatitis, alopecia and diarrhea. It occurs in one of two forms: an inborn (congenital) form and an acquired form.

A consecutive case series of 5 patients with ADE were presented.

All the 5 cases had rashes in the periorificial areas, diarrhea and low serum zinc levels. Two brothers were among the cases seen. ADE is not uncommon in Nigeria, both clinical variants are encountered.

Key words: acrodermatitis; enterohepathica, cases, Nigeria

INTRODUCTION

Acrodermatitis enterohepatica is a rare bullous skin disorder. It is either inherited recessively or acquired. It is characterized by diarrhea, rash around the mouth and/or anus and alopecia. Both males and females are equally affected. Symptoms usually occur in bottle fed infants within weeks after birth or breast fed infants soon after weaning.

METHOD

This is a consecutive case series of 5 patients with acrodermatitis enterohepathica seen in dermatology clinic of Aminu Kano Teaching Hospital. All patients were clinically diagnosed and confirmed with serum zinc level measurement.

RESULT

A total of 5 patients were seen .The epidemio-clinical characteristics is summarized in Table 1. All the 5 patients were males. They are all aged between 1 and 3years.One had the disorder since first year of life, and it appeared in the other 4 patients in the second year of life, among who are two siblings. All the cases presented with peri-orofacial, acral and genital dermatitis, irritability and diarrhea. The inherited form of AE was observed in 2 of the patients who are product of consanguineous marriage (Fig. 1a & b, Fig. 2a & b). The other three cases (Fig. 3, 4, 5) presented with classical acquired form of the disease. Anthropometric measurement showed one of the patients to be undernourished with z-score lower than 2 standard deviations than expected for age and sex using the WHO standard growth chart.

Another patient had severe acute malnutrition with z-score less than 3 standard deviations than what is expected for age and sex according to standard WHO growth charts. While the remaining three were well nourished with z-score above 2 standard deviations expected for age and sex. Systemic examinations were not remarkable in all the patients.

Serum zinc level in all the five patients was less than 60µg/dl (normal 60-140µg/dl).

The five patients were successfully treated with oral zinc therapy. Those patients with evidence of malnutrition were co-managed with nutritionist for nutritional rehabilitation.

DISCUSSION

Acrodermatitis enterohepathica is a rare form of zinc metabolism. It can occur as an inborn form inherited in autosomal recessive pattern that occur as a result of intestinal abnormalities that lead to in

ability to absorb zinc from the intestine.[1] It also can be in an acquired form due to inability of the mother to secrete zinc into her breast milk because of genetic abnormality in the mother due to single mutation in the SLC30A2 gene where zinc-binding co-factor produced by the pancreas is absent, or as a result of by-pass surgery in the child, removing part of upper intestine where absorption of zinc occur. It can also be from inappropriate amounts of zinc in special intravenous nutritional preparations, or from reduced dietary intake of zinc.[1,2] Both inborn inherited and acquired forms are characterized by peri-orificial and acral dermatitis, diarrhea, and alopecia.[1,2.] The exact cause of the inherited recessive form is not known but may be related to mutation in a gene (SLC39A40) that codes for the zinc transporter protein ZIP4 and the missing protein may be responsible for decrease zinc uptake and its abnormal metabolism.[3]

The role of zinc in the treatment of Acrodermatitis enterohepathica was discovered in 1973, before then the inherited form of the disease was usually fatal. With treatment, all patients with Acrodermatitis enterohepathica can lead normal lives.

In this case series, two of the cases had the inherited form and had a family history, while the remaining three cases were mainly acquired and occurred after weaning from breast milk to foods with in adequate zinc content. The perioral area is the site reported to be most commonly affected, followed by anogenital area. Other features include conjunctivitis, sensitivity to light, loss of appetite, diarrhea, mild or severe irritability, depressed mood, growth failure, alopecia and nail changes (paronychia and nail dystrophy).[5]

Among our five cases it was observed that perioral and anogenital area was the commonest site affected as all the cases presented with it. Two of the cases presented with features of protein energy malnutrition and all the five cases were apathetic and irritable on presentation. One of the cases presented with nail dystrophy and two presented with alopecia. All the five cases presented with recurrent diarrhea.

Clinically the differential diagnosis of Acrodermatitis enterohepathica may include seborrheoic dermatitis, psoriasis, kwashiorkor, and marasmus, Celiec sprue.[6,7,8] Low plasma zinc level was observed in all the five cases.

Acrodermatitis enterohepathica may requirelifelong Zinc supplementation.[4] Genetic counseling is recommended for families of patients with the congenital form of AE.[9]

All the five cases we reported showed significant improvement within 2-3weeks of zinc supplementation.

AGE (YEARS)	TIME OF ONSET OF SYMPTOMS	SERUM ZINC LEVEL	PRESENTATION/S
2	8months	50µmol/dl	Periorifacial and anogenital dermatitis,diarrhea.irritability
3	6months	38µmol/dl	Periorifacial and anogenital dermatitis,diarrhea,irritability.
2	18months	42µmol/dl	periorifacial, acral and anogenital dermatitis, growth failure,irritability
18months	6months	34µmol/dl	Periorifacial, acral and anogenital dermatitis,irritability,growth failure,diarrhea
3	2years	52µmol/dl	Periorifacial and acral dermatitis, irritability,diarrhea, nail dystrophy

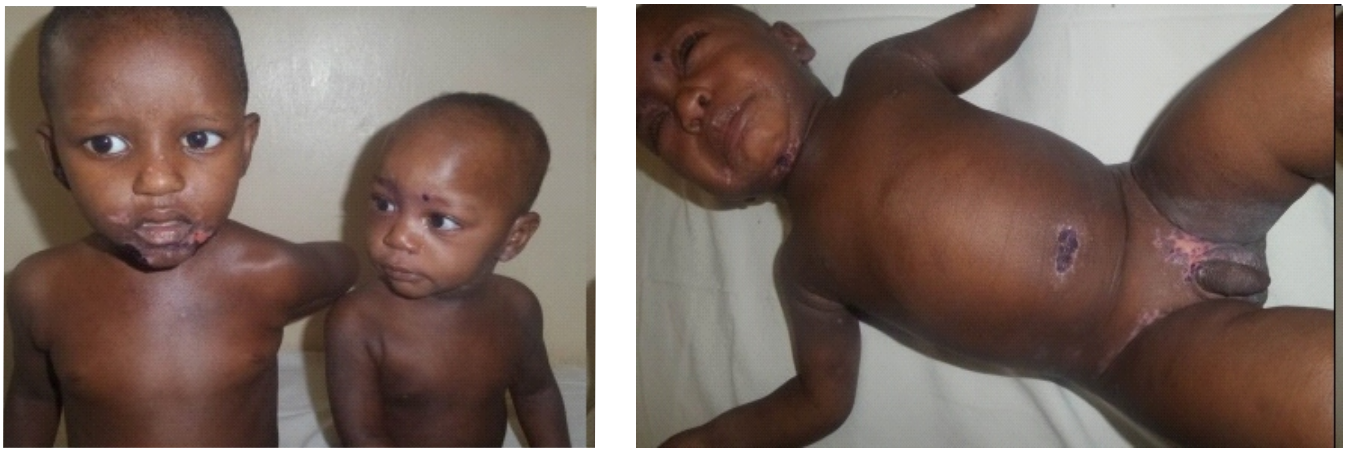


Fig 1. The two siblings with inherited form of ADE before treatment

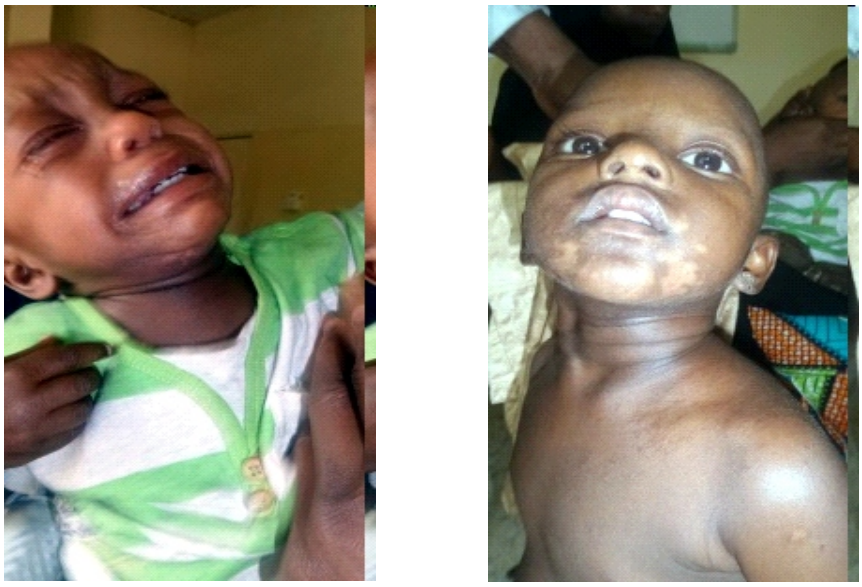


Fig. 2: The inherited form in siblings 2weeks after commencement of zinc therapy

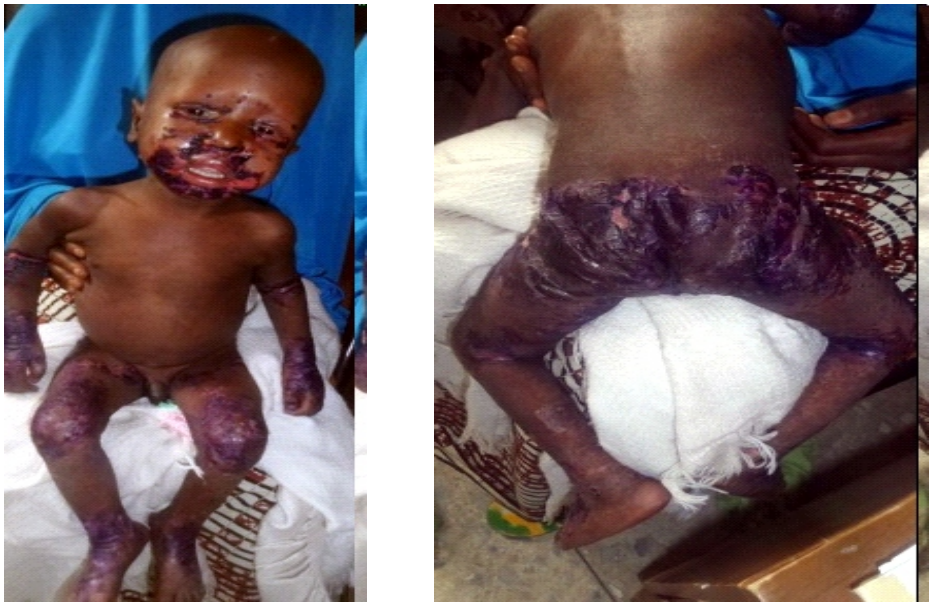
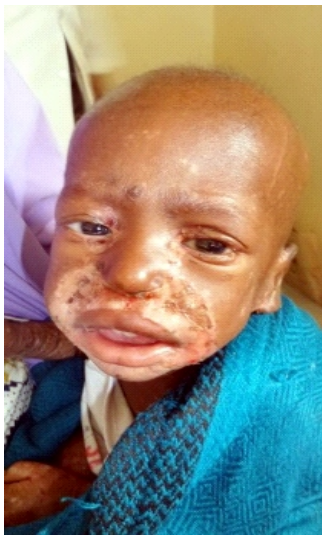


Fig. 3a: Case 3 after 2 weeks on treatment



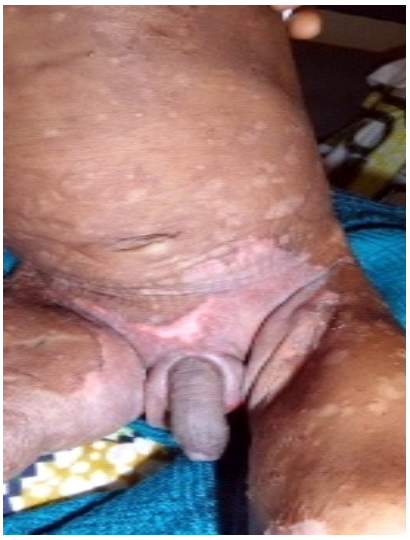
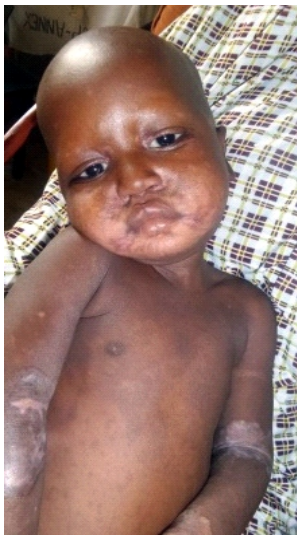


Fig. 3b: Case 3 after 2 weeks on treatment



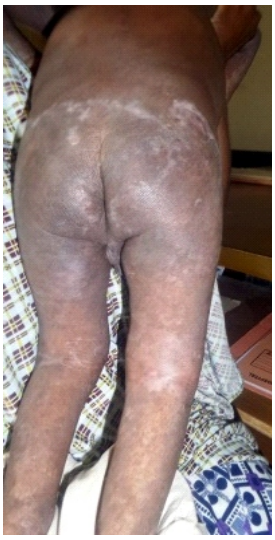


Fig. 4: Case 4

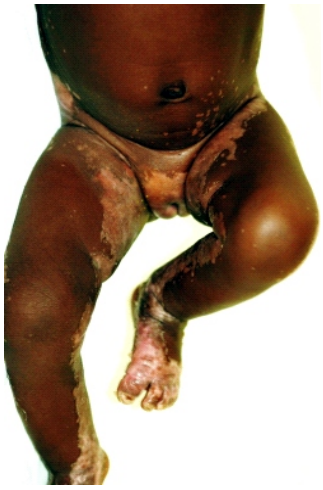


Fig. 5: Case 5

CONCLUSION

Acrodermatitis enterohepathica is not uncommon in Nigeria. Oral zinc is cheap and effective mode of treatment alongside other measures.

REFERENCES

1. Lee SY, Jung YJ, Oh TH, Choi RH. A case of Acrodermatitis enterohepathica localized on Hands and feet with a Normal serum zinc level. AnnDermatol 2011; 23:88-90.
2. Perafan-Riveros C, Franca LF, Alves AC, Sandes JA. Acrodermatitis enteroheptica. Case report and review of the literature. PediatrDermatol 2002;19:426-31.
3. Kury S, Dreno B, Brezieau S. Identification of SLC39A4, a gene involved in acrodermatitis enterohepathica. Nat Genet 2003; 31:239-40.
4. Ngan V. Acrodermatitis enterohepatica. 2004. Derm Net NZ.
5. Van Wouwe JP. Clinical and laboratory diagnosis of acrodermatitis enterohepathica. Am J Dermatopathol 1982; 4:303-11.
6. Brandt T. Dermatitis in children with disturbances of general condition and absorption of food. ActaDermatol Venerolica1936; 17:513-546.
7. Abrams S. Zinc deficiency and supplementation in children and adolescents. Update.
8. Selgal VN, Jain S. Acrodermatitis enterohepathica. ClinDermatol 2000; 18:745-8.
9. Radja N, Charles-Holmes R. Acrodermatitis enterohepathica lifelong follow-up and zinc monitoring. ClinExpDermatol 2002; 62-3.