Case Report

Acrodermatitis Enteropathica In Adolescence — A Rare Presentation of Malabsorption

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Acrodermatitis Enteropathica (AE) is a rare, inherited or acquired disorder due to Zinc Deficiency. It is characterized by a triad of Acral Dermatitis, Alopecia and Diarrhoea. The acral and periorificial distribution of the rash is considered as a pathognomonic cutaneous marker for Zinc Deficiency. It is inherited as an autosomal recessive disorder and usually manifests in weaning children. Acquired cases can manifest at any age and occur secondary to malabsorption, dietary insufficiency, pancreatic insufficiency, chronic alcoholism etc. Supplementation with Zinc produces excellent clinical improvement and reduces mortality.

In our case, we diagnosed an adolescent girl with AE secondary to malabsorption who presented with periorificial ulcers, chronic diarrhoea and alopecia.

This case highlights the importance of keeping a low threshold for suspicion of Zinc Deficiency in patients with such skin manifestation. Otherwise the patient might be misdiagnosed as skin infection and thus not respond to treatment. Prompt diagnosis facilitates proper therapy and complete recovery in a condition which can be debilitating and fatal if not treated.

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Key words **: Acrodermatitis Enteropathica, Zinc, Periorificial, Alopecia, Diarrhoea, Malabsorption.**

Swedish dermatologist Thore Brandt first described a case of severe Zinc Deficiency in 1936 and thus named it as Brandt's Syndrome, also known as Acrodermatitis Enteropathica. It may be inherited or acquired. We report here a case of acquired Acrodermatitis Enteropathica in an adolescent girl.

Case Report

A 13 year female hailing from a village in Silchar presented to Medicine OPD with complaints of erythematous scaly & crusting lesions at multiple sites, chronic diarrhoea, easy fatiguability and lethargy for the last 1year.The skin lesions initially started around the perineal region, tips of fingers & toes as blackish discolouration & itching. It gradually progressed and there was excoriation, reddening & crust formation. With time, the lesions started appearing around her eyes & mouth. The patient had visited several local physicians who treated her as a case of Cellulitis.She also took many over the counter medications for her skin lesions but it did not improve.

There was no history of any diagnosed hereditary disease in the family. There was no H/O similar complaints in the family.There is a H/O sibling death around 1month after birth but the exact cause of death is not known. At present, the patient has 2 siblings who are in good health.

Editor's Comment :

- This case highlights the importance of a correct diagnosis in treating a patient with a debilitating disease and the satisfying result of prompt intervention.
- The threshold for suspicion of zinc deficiency in patients with such skin manifestation should be low. Otherwise the patient might be misdiagnosed as skin infection and thus not respond to treatment.

On examination, patient was afebrile, conscious, oriented & irritable. Built was asthenic, BMI=14.7kg/m^2. Pallor, bilateral pitting oedema and ascitis were present. Periocular, perioral, perineal & acral lesions with desquamation & crusting were seen (Fig 1).Bilateral ectropion was present. Hair was scanty and coarse with brown discoloration. (Fig 2).Other systemic examination were not significant.

Investigation revealed a microcytic hypochromic anaemia, Red Cell Indices showed (WBC-5.41*10^3/µL, RBC-2.82*10v /µL, Hb-6.9g/dL, HCT-25.7%, MCV-91.1fL, MCH-28pg, MCHC-30.7g/dL, PLT-310*10^3/µL, RDW-CV-14.1%, MPV-10.9fL, DLC. Neutrophil-76.4%;Lymphocyte-20.6%;Monocyte-2.7%;Basophil-0.3%;Eosinophil-0%), hypoalbuminaemia, hypokalemia, and hypochloremia & iron deficiency. Serum Zinc level was 15ug/dL. Ultrasonography of the abdomen was normal except for moderate ascitis. Ascitic fluid showed (Protein-1.6g/dL,

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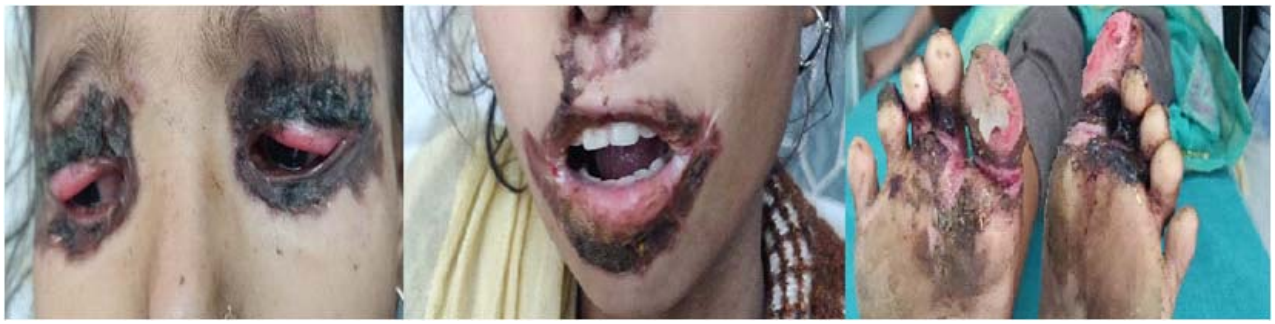

Fig 1 — Periorificial & acral lesions at presentation

Glucose-78mg/dL, Total cell count-6/µL, RBC-0, WBC-6/ µL, PMN-4/ µL, MN-2/ µL, SAAG-1.3). Stool routine examination was normal and stool culture was sterile. Other investigations including autoimmune markers, anti tTG were non reactive.

On the basis of history, examination & investigations we came to the diagnosis of Acrodermatitis Enteropathica secondary to malabsorption. The patient was supplemented with Oral Zinc Acetate Tablets 50mg twice daily along with ORS, probiotic therapy,vitamin A, Iron Folic acid supplementation and Doxycycline 100mg twice daily for 2 weeks. Diet rich in protein, vitamins and minerals was advised. Local application of topical corticosteroids was adviced.She also received 1 unit of packed RBC transfusion.

By 2 weeks patient improved dramatically. The skin lesions cleared up and the diarrhoea subsided. General well being of the patient also improved (Fig 3).

Fig 2 — Scanty & coarse hair with brown discolouration

Discussion

Inherited Acrodermatitis Enteropathica is a rare autosomal recessive disorder. It usually begins within days to weeks after birth in infants bottle fed with animal milk or soon after weaning from the breast in older infants[1]. Acquired Acrodermatitis Enteropathica usually occurs in patients receiving prolonged total parenteral nutrition, or secondary to chronic alcoholism, pancreatic or dietary insufficiency and malabsorption.The clinical syndrome consists of a triad of acral dermatitis, alopecia and diarrhoea. The acral and periorificial lesions are in fact pathognomonic cutaneous marker for Zinc deficiency. Other clinical features include apathy, irritability, growth retardation, failure to thrive, stomatitis and delayed wound healing. Response to Zinc therapy confirms the diagnosis[2].

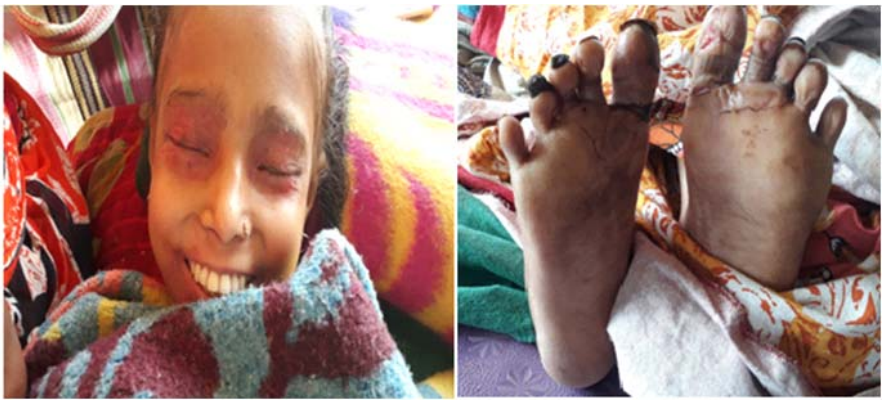

Fig 3 — Improvement of the patient by 2 weeks

Zinc is an important constituent of the human body because it forms an integral part of many metalloenzymes.Unmilled cereals, beans, cheese, whole wheat bread, meat, shellfish, nuts and legumes are good sources of bioavailable Zinc[2]. The requirement of Zinc for adolescent male and female are 15mg and 12mg /day respectively[1]. The supplementation of Zinc

sulphate for Acrodermatitis Enteropathica was first introduced in 1973[3-5]. Dosage is based on the amount of elemental Zinc present in the preparation. As observed in a study treatment with Zinc Sulphate 1mg/kg/day helped clear the symptoms in 5 days[6]. In yet another case report, Zinc Sulphate was used in a dosage of 5mg/kg/day[7]. There was rapid improvement of diarrhoea within 24 hours and the skin lesions within 1 to 2 weeks[8].

Our case highlights the importance of a correct diagnosis in treating a patient with a debilitating disease and the satisfying result of prompt intervention. The threshold for suspicion of Zinc deficiency in patients with such skin manifestation should be low. Otherwise the patient might be misdiagnosed as skin infection and thus not respond to treatment.

The exact cause for malabsorption could not be found out.Ultrasonography of abdomen was normal except for moderate ascitis.Her stool examination was normal and stool culture showed commensal flora of gastrointestinal tract.Our patient was advised for endoscopic intestinal biopsy but the attendants refused to give their consent for the procedure.

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