DOI: 10.21276/IJIPSR.2018.06.12.761

International Journal of Innovative Pharmaceutical Sciences and Research
www.ijipsr.com

ACRODERMATITIS ENTEROPATHICA – A CASE REPORT

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Abstract

Acrodermatitis Enteropathica (ADE) is a rare genetic inherited or acquired autosomal recessive disorder, characterized by acral and periorificial dermatitis, alopecia, and diarrhea which is caused by mutations in the gene that encodes a membrane protein that binds zinc and further leads to impaired absorption of zinc from the gastrointestinal tract. Early recognition and treatment of the disease will decrease morbidity and mortality. The preferential involvement of acral and peri-orificial skin is a feature that is pathognomonic for zinc deficiency. The incidence is about 1 in 500,000 and the condition affects without predilection of race and sex. We present a case of acrodermatitis enteropathica a 40 yr old male with zinc deficiency from childhood.

Keywords: Acrodermatitis Enteropathica (ADE), autosomal recessive disorder, acral and periorificial dermatitis.

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DOI: 10.21276/IJIPSR.2018.06.12.761

INTRODUCTION

Acrodermatitis Enteropathica is the deficiency of zinc arised from acquired or genetic causes in which acquired zinc deficiency develops from lack of zinc in the diet or decreased uptake of zinc in the duodenum or jejunum where as the genetic zinc deficiency can be traced to a mutation in the gene responsible for zinc transportation, resulting in poor zinc absorption [1]. Zinc is an essential mineral that has been found to be present in at least 100 metalloenzymes wherein zinc deficiency, whether a result of an acquired or inherited abnormality is associated with characteristic cutaneous findings [2]. The ideal clinical manifestations of acrodermatitis enteropathica are distinguished by the triad eczematous and erosive dermatitis, acral and perioficial symmetrical dermatitis, alopecia and diarrhea. Paronychia, onicodistrophy, angular stomatitis, cheilitis, conjunctivitis and photophobia can also take place and the disorder progresses with difficulties regarding- weight gain, delayed growth, neuro-psychic disorders, delayed puberty, male hypogonadism, anemia, anorexia, hipogeusia and difficulty to heal wounds. Despite of the cause, zinc deficiency changes immunity, contributing with the high predisposition to fungal and bacterial infection which can trigger systemic severe scenarios [3]. Identification of this condition is important as it can be fatal in severe cases and as it occurs due to impaired Zinc absorption and patients respond dramatically when treated with Zinc [4].

CASE PRESENTATION

A 40yr old male patient presented with the complaints of painful skin lesions since 3months, developed crusted lesions associated with redness initially over both feet which is gradual in onset and progressed to involve knees, genitals, axilla, face, scalp, neck, back.

He gave the history of photosensitivity, excessive loss of hair, watery discharge from eyes, burning sensation, photophobia since 3months. There was no history of diarrhea. He gave the history of Zinc deficiency since childhood and on Zinc supplements (T. Ascozin 50mg TID) since childhood. He experienced similar complaints of skin manifestations 1yr back. He was on irregular zinc supplementation from one and a half year and completely stopped taking medications since 6months. He follows a mixed diet and has a habit of smoking since 15yrs. His family history includes consanginous marriage of his parents, similar complaints in the younger sister and died at the age of 15. Rest of his siblings were normal.

On clinical examination multiple crusted erosions were seen over face, scalp, neck, palms, dorsum of hands, extensor aspect of both elbows, axilla, groin, both knees, dorsal aspects of both

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feet extending onto legs associated with multiple scaly popular lesions seen over chest and back. There was absence of eyelids and eyebrows, oral cavity thrush was seen, nails were found to be normal.

Fig.1

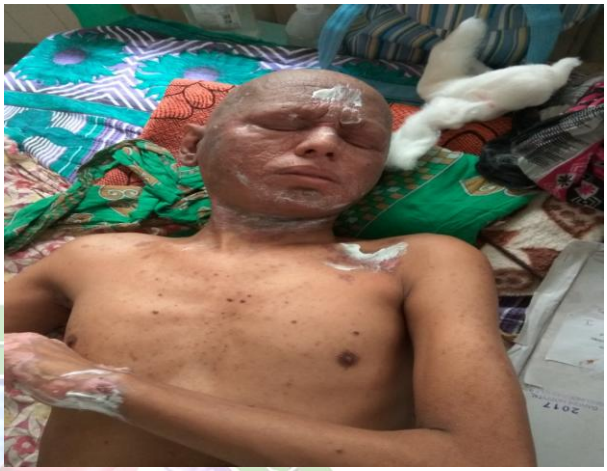

Fig.2

Fig.1: Periorificial erosive dermatitis associated with alopecia on the day of admission.

Fig.2: Healed skin after 12days of therapy.

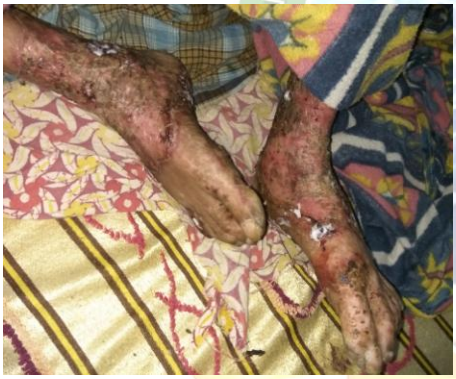

Fig. 3

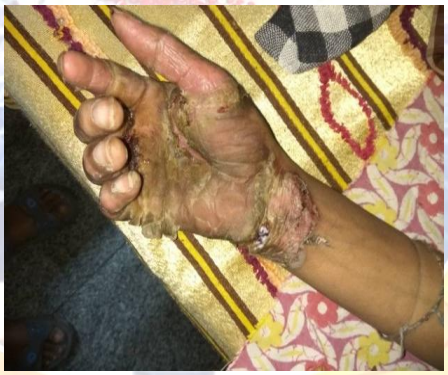

Fig. 4

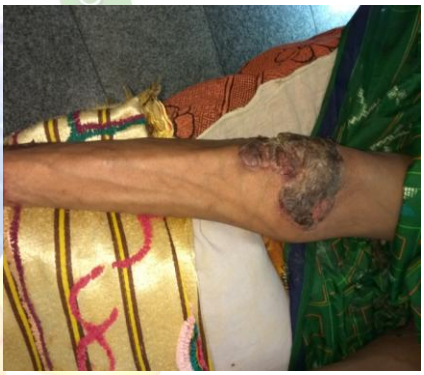

Fig. 5

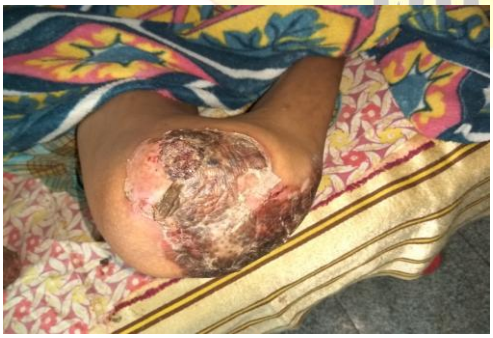

Fig. 6

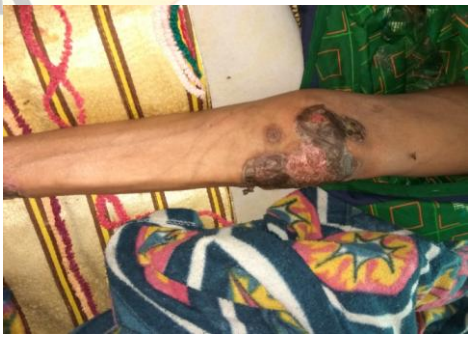

Fig.7

Fig.3,4: Acral erosive dermatitis.
Fig.5,6,7 : Crusted erosions on the extensor parts of elbows and knees.

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His laboratory investigations revealed decreased serum zinc 45microgm/dl(60-120micromg), raised erythrocyte sedimentation rate 30mm/hr, decreased alkaline phosphatase 29IU/L (44-147IU/L), increased SGPT 131IU/L (7-56IU/L), decreased serum albumin 2.3gm/dl (3.5-5gm/dl). Other LFT and RFT parameters, blood sugar, CBP were within normal limits.

Patient has been treated with

1. Elemental zinc- Tab. Ascazin 50mg TID
2. Multivitamin- Tab. Zincovit OD
3. Chitomesh-SF cream for E/A BD
4. Liquid paraffin oil for E/A BD
5. Framycetin Sulphate BD
6. Saline Compression
7. Tab. Flucanazole 150mg OD for every 3days
8. Inj. Piptaz 4.5mg IV BD
9. Inj. Pantoprazole 40mg IV OD
10. Lubrex E/D
11. Lacrigel eye ointment at bed time
12. Ciplox E/D

He was discharged after 15days after he got relieved from the condition by prescribing oral zinc supplements Tab. Ascozin 50mg thrice a day on a regular basis. Patient was conselled about the importance of taking medication regularly and its effectiveness in preventing the recurrence of the condition.

DISCUSSION

Zinc is an crucial trace element essential for the proper functioning of all cells, and plays an vital role in the metabolism of protein, carbohydrate, and vitamin A and is a cofactor of numerous metal enzymes such as alkaline phosphatases, alcohol dehydrogenase, RNA polymerase, and numerous digestive enzymes. Zinc deficiency can be acquired or inherited wherein the causes of acquired zinc deficiency: premature infants, low birth weight, zinc deficiency in maternal milk, exclusive parenteral nutrition, malabsorption syndromes such as Crohn disease and celiac disease, alcoholism, low calcium and phytate diet, and kwashiorkor and the hereditary deficiency of zinc classically known as " acrodermatitis enteropathica." which was first described in 1936, is a rare autosomal recessive mutation of SLC39A4 (solute carrier family 39 member 4) gene on

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chromosome 8q24.3, which determines a congenital partial or total deficiency of the zinc transporter protein zinc-ligand binding protein 4 (ZIP 4) Acrodermatitis enteropathica occurs at an estimated incidence of 1 per 500,000 children worldwide with no preference for race or gender. In developing countries such as Southeast Asia and sub-Saharan Africa, zinc deficiency can be seen in a third of the population and in developed countries such as the United States, inadequate zinc intake can be seen in vegetarians, alcoholics, malnourished, and premature infants [1,5].

The three dominant clinical signs of Acrodermatitis enteropathica are lesions with localization around the body orifices and on the extremities, total alopecia, and diarrhea in which diarrhea may be mild or severe, present with fatty substances in the feces (steatorrhea) and in the congenital form symptoms start gradually, frequently at the time of weaning of an infant; skin around body openings such as the mouth, anus, and eyes, and the skin on elbows, knees, hands, and feet become inflamed and the skin lesions are usually blistered (vesicobullous), after drying out become psoriasis-like where as the skin around the nails may also be inflamed and the nail may be abnormal due to malnourished tissue. Hair loss on the scalp, eyelids, and eyebrows may be total (alopecia) and usually occurs along with the Inflammation of the membrane that lines the eyelid (conjunctivitis) [6,7].

Initial assessment of a patient's zinc status is determination of plasma or serum zinc concentrations (plasma zinc concentrations are preferred). Blood should be drawn before breakfast and the patient should not receive zinc supplements on the day of the test. A second approach to determining body zinc status is to measure the activity or concentration of zinc-dependent enzymes, but there is yet no consensus on which zinc-dependent enzyme is most appropriate to measure [7].

Zinc sulphate for acrodermatitis enteropathica was first introduced in 1973. Oral zinc given in a dose of about 2mg/kg/day was found to cure all clinical manifestations related to zinc deficiency within 1 or 2 weeks. In most instances dietary supplementation with two to three times the RDA in doses of 30 to 55mg of elemental zinc daily is adequate to restore normal zinc status [8]. Treatment includes supplementation of elemental zinc at a dose of 2 mg/ kg/day, at least two or three times the recommended dietary allowance of 15 mg/day.[(4)] Most authors recommend an initial dose of 5-10 mg/kg/d with maintenance doses of 1-2 mg/kg/d and AE must be treated and monitored throughout the entire life. In the event of treatment interruption, relapses are inevitable and the cutaneous signs are the first to recur. The long-term prognosis is excellent [9].

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CONCLUSION

This case emphasizes the need for regular zinc supplementation through diet and medications that is required to reverse the condition, reduce mortality and prevent the long term consequences of zinc deficiency. A review from a dietician is very beneficial in ensuring patients have adequate zinc consumption.

ACKNOWLEDGEMENT

We express our profound gratitude to all the doctors of In-patient department of dermatology for providing unflinching support.

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