

**CASE REPORT****A RARE CASE OF SEVERE ACRODERMATITIS ENTEROPATHICA DURING COVID-19 LOCKDOWN**

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Acrodermatitis Enteropathica is a rare hereditary condition characterized by perioral dermatitis, diarrhoea, and alopecia. Aetiology includes autosomal recessive inheritance and acquired causes such as protein malnutrition, malabsorption syndromes, premature births, parenteral nutrition, chronic illnesses, and alcoholism. We report a rare case of a 12-year-old boy who presented with cutaneous manifestations involving the whole body, diarrhoea, and sparse hairs. A low level of plasma zinc, and alkaline phosphatase level was found, and he was started on zinc supplements. He significantly improved in a few days, and was discharged after counselling the parents about treatment compliance. This case highlights importance of early diagnosis, and the importance of treatment compliance in Acrodermatitis Enteropathica. It also stresses the need to take measures to ensure the provision of health facilities especially in remote areas.

**Keywords:** Acrodermatitis enteropathica; COVID-19 Lockdown; Treatment non compliance

**Citation:** Hussain T, Kumar J, Aslam T, Khan S, Ishaq M, Kumar D. A rare case of severe Acrodermatitis Enteropathica during COVID-19 lockdown. J Ayub Med Coll Abbottabad 2022;34(4):880–2.

**DOI:** 10.55519/JAMC-04-10371

**INTRODUCTION**

Acrodermatitis Enteropathica is caused by severe zinc deficiency due to a defect in gene SLC39A4 encoding a protein that helps in zinc absorption.<sup>1</sup> It has an incidence of 1/500,000 births and has no relation to gender and ethnicity.<sup>2</sup> The clinical manifestations in Acrodermatitis enteropathica vary with the age. However, the important manifestations include diarrhoea, skin lesions, alopecia, and infections.<sup>3</sup> Zinc is an essential part of many metalloenzymes, which helps in cellular metabolism.

The diagnosis of acrodermatitis enteropathica is based on clinical findings and laboratory investigations including plasma zinc and alkaline phosphatase level which indirectly helps in diagnosis. However, genetic testing is done for confirmation but it is not routinely performed. The treatment involves supplementation of zinc mostly in the form of zinc sulfate. In this case the 12-year-old boy had a recurrence of disease due to non-compliance to treatment. The patient lived in a remote area and could not get his medicines due to Covid-19 imposed lockdowns. After restarting treatment, he showed remarkable response to zinc supplements.

**CASE PRESENTATION**

A 12-year-old boy presented with diarrhoea and cutaneous lesions involving the face, upper limbs, lower limbs, and buttock. He was vitally stable. On examination multiple crusted, eroded, erythematous papule, and plaque were noted on perioral facial areas, symmetrically on bilateral upper limbs, acral part of lower limbs, and buttock region as shown in the (Figure-1, 2, and 3). Alopecia was also seen along with oral thrush. According to the parents of the patient, he had this condition since he was one year old. His diet was normal and weaning was initiated at 6 months.

One of his siblings had similar dermatologic manifestations. He did not have any features of malabsorption syndromes. They had numerous dermatologic consultations. He was previously diagnosed as acrodermatitis enteropathica based on lab tests of zinc level and was on the maintenance dose of zinc sulphate according to his age. However, recently due to the lockdown imposed by the government during the covid-19 pandemic, they were unable to continue his treatment which aggravated his condition.



**Figure-1 and Figure-2: multiple crusted, eroded, erythematous papule and plaque symmetrically on bilateral upper limbs, and on perioral facials areas**



**Figure-3: Multiple crusted, eroded, erythematous papule and plaque symmetrically on bilateral lower limbs.**



**Figure-4: Hands after treatment with zinc**



**Figure-5: Face after treatment with zinc**

The laboratory investigations were sent including zinc level and the entire baseline. All other labs including complete blood count, liver function tests, urea, creatinine, anti-tTG and IgA anti endomysial antibodies were found normal, apart from zinc level of 24ug/dl and alkaline phosphatase was 40 IU/L, which supported our diagnosis. Genetic test could not be done due to non-availability and non-affordability of testing abroad. Although at first sight of the patient, we had many differential diagnoses such as seborrheic dermatitis, atopic eczema, pellagra, psoriasis, and Langerhans cell histiocytosis, but they were all ruled out by history, physical examination, and lab findings. The patient was initiated zinc supplements in the form of zinc sulphate. His condition improved dramatically within 3–4 day as shown in (Figure-4, 5). Furthermore, a follow-up visit was scheduled.

## DISCUSSION

Zinc is one of the essential micronutrients which have a vital role in the body functioning such as immune response, development, wound healing, and metabolism.<sup>4</sup> Zinc deficiency can be hereditary or acquired.

The hereditary cause of zinc deficiency (Acrodermatitis Enteropathica) is because of mutation in gene SLC39A4 located at 8q24.3 chromosome.<sup>5</sup> Which encode for Zip4 protein and assist in zinc transport across the intestinal lumen.<sup>6</sup> The acquired causes include protein malnutrition such as Marasmus and Kwashiorkor, malabsorption syndromes including Celiac disease and Crohn disease, premature births, parenteral nutrition, chronic illnesses, and alcoholism.<sup>7</sup>

The clinical presentation of zinc deficiency encompasses skin manifestations (bullous-pustular dermatitis, atopic dermatitis), GI-symptoms (diarrhoea), hair abnormality (alopecia, hair loss), neuropsychiatric problems (depression, altered behaviour), delayed puberty, growth, and mental retardation.<sup>8</sup> However, the classic clinical trial in Acrodermatitis Enteropathica includes diarrhoea, dermatitis, and alopecia. In our case, the patient presented with a similar clinical trial of diarrhoea, dermatitis, and alopecia.

The diagnosis of Acrodermatitis Enteropathica is based on characteristic clinical findings most importantly morphology of cutaneous lesions, zinc level in plasma, and the response to treatment, which has great significance especially in resources limited areas.<sup>9</sup> In our

case, the patient showed a dramatic response to treatment in a few days. Moreover, a biopsy of skin lesion helps in supporting diagnosing and rules out other differential diagnosis.<sup>10</sup> The management of Acrodermatitis Enteropathica involves lifelong supplementation of zinc in the form of zinc sulphate, acetate, or gluconate. It is considered that zinc in the form of zinc sulphate is preferred and more tolerable. The recommended dose is 1–3 mg/kg/day.<sup>11</sup> Compliance with treatment is very essential because it prevents recurrence, and resolves the features associated with Acrodermatitis Enteropathica, and restores immune function.<sup>12</sup>

**Consent:** Consent was taken from the guardian of the patient.

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Submitted: November 11, 2021

Revised: December 14, 2021

Accepted: March 27, 2022

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