Acquired Acrodermatitis Enteropathica: A Case Report

L Kuhnel[1], W Hong[1], M Smith[1], A Sleem DO[2] , K Barron PharmD[3], T Nguyen PharmD[3], P Becnel MD[2], M Marshal MD[2]

[1]Faculty of Medicine, University of Queensland-Ochsner Clinical School, New Orleans, LA

[2]Department of Hospital Medicine, Ochsner Medical Center, New Orleans, LA

[3]Department of Pharmacology, Ochsner Medical Center, New Orleans, LA

Introduction

- Acrodermatitis enteropathica (AE) is a rare dermatologic disorder associated with zinc deficiency.
- Acquired AE has no definitive diagnosis to date.[2] Usually clinical suspicion coupled with low zinc findings are used to make the diagnosis.[1] However, zinc levels in serum, urine, or hair, have not been found to be specific or sensitive.[3] A skin biopsy is also not specific.
- We present the case of a 49 year old African American female with past medical history of alcoholic liver cirrhosis and chronic kidney disease stage 5. Her family history was unremarkable.
- She was brought to the emergency department with complaints of confusion and exacerbation of diffuse desquamating, crusting skin lesions extending from the palms and soles to the dorsum of extremities, axillae and abdomen while sparing the face and oral mucosa.
- During a previous hospitalization, patient was noted to have low plasma zinc levels. She was given oral zinc therapy, to which she was not responsive.

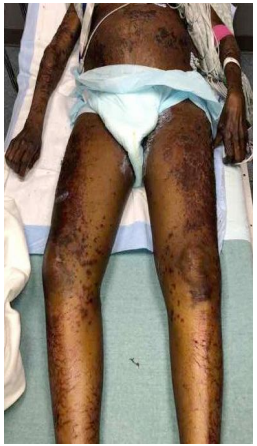

Figure 1: Patients rash noted on day one of hospitalization: diffuse desquamating, crusting skin lesions extending from the palms and soles to the dorsum of extremities, axillae and abdomen and sparing the face and oral mucosa

Hospital Course

- A diagnosis of acquired AE was previously suggested based on low plasma zinc values noted during a prior hospitalization.
- Further workup included a skin biopsy, results of which, while not confirmatory, were consistent with AE.
- A 5 day trial of IV zinc 20 mg in PPN form (3 mg/kg dosing) in the hospital setting was initiated.
- Patient was switched to oral zinc supplementation after the trial was completed.
- Improvements in the skin were first noted on day 3.
- After completion of the trial of parenteral zinc, the patient's skin showed significant improvement.
- These improvements continued until the patient expired due to a transfusion-related acute lung injury (TRALI).

Discussion

- The acquired form of AE is less studied and presents with more diagnostic uncertainty then its genetic primary counterpart.
- Treatment for both types of AE studied in previous literature has indicated for oral zinc supplementation. Our patient did not respond to oral zinc therapy initiated after her first hospitalization.
- We confirmed the patient's previous AE diagnosis based on the response to parenteral zinc supplementation.
- We postulate that her original zinc deficiency and lack of response to oral zinc was malabsorptive in nature and may be associated with patient's history of cirrhosis.

Conclusions

- Acrodermatitis enteropathica, is a rare condition, mostly seen in infants as primary AE as acquired AE is less seen and poorly understood. Insight into a patient's zinc levels can help guide diagnosis and management.
- However, zinc correction in acquired AE can be challenging as the etiology of the zinc deficiency must be known to intact sufficient treatment.
- In this case, malabsorption and chronic malnutrition from advanced liver disease was the cause of the manifestation of this rare condition leading to trial of slightly unconventional management techniques for treatment, that proved to be successful.

References

1. Schmitt S, Küry S, Giraud M, Dréno B, Kharfi M, Bézieau S. An update on mutations of the SLC39A4 gene in acrodermatitis enteropathica. Hum Mutat. 2009 Jun;30(6):926-33. doi: 10.1002/humu.20988. PMID: 19370757.
2. Livingstone C. Zinc: physiology, deficiency, and parenteral nutrition. Nutr Clin Pract. 2015 Jun;30(3):371-82. doi: 10.1177/0884533615570376. Epub 2015 Feb 13. PMID: 25681484.
3. Nistor N, Ciontu L, Frasinariu O-E, Lupu VV, Ignat A, Streanga V. Acrodermatitis enteropathica: A case report. Medicine (Baltimore). 2016;95(20):e3553.