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Acquired Acrodermatitis Enteropathica Presenting as Confusion and Desquamating Maculopapular Rash

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Background

- Zinc is an essential micronutrient in maintaining skin integrity, wound healing, and immune function.
- Zinc deficiency can cause the clinical condition acrodermatitis enteropathica (AE), and may present with the characteristic erythematous scaly rash, alopecia, diarrhea, mental status changes, depression, and immune dysfunction.
- In AE, skin manifestations may be accompanied by alopecia and diarrhea, but this classic triad is present in only 20% of cases.
- We present a case of acquired AE presenting with skin findings, confusion and diarrhea.

Case Presentation

- A 56-year-old female presented to the hospital with confusion, diffuse rash, and loose stools from a nursing facility that she was placed about one month before admission.
- Prior to staying at a nursing facility, she was living in an abusive household with limited access to food and had not seen a physician for more than 30 years.
- The skin examination revealed a pruritic, diffuse maculopapular rash with desquamation on the cheeks, chest, upper and lower extremities including the palms and feet. (Image 1). Skin examination also demonstrated perioral fissuring and paronychia of all fingers.
- BMI on admission was 17.2 and blood pressure was 80/50 mmHg.



Figure 1. Generalized erythematous patches and plaques with desquamation on both arms

Clinical Course

Pertinent laboratory findings on initial admission:

K: 3.3 mmol/L (3.5 - 5.2 mmol/L)

Na: 134 (135 - 145 mmol/L)

Creatinine: 2.46 mg/dL (0.50 - 1.10 mg/dL)

25-hydroxyvitamin-D: 5 ng/mL (30 -100 ng/mL)

Zinc: 55 ug/dL (60-130 ug/dL)

Glucagon: 93 pg/mL (<80 pg/mL)

- Work-up for infectious and malabsorptive causes including celiac disease were negative.

Treatment: Aggressive supplementation of appropriate vitamins, IVF, and Zinc sulfate 220 mg BID was started.

Follow-up: 8 weeks after treatment initiation, Zinc level was increased to 156 ug/dL.

- Rash had completely resolved, she was no longer experiencing diarrhea or cognitive dysfunction.
- Patient's BMI has improved to 20.6
- Zinc sulfate was stopped and multivitamins containing zinc was continued.

Discussion

- Many of the clinical features of AE are nonspecific.
- A high index of suspicion is needed especially in the appropriate clinical context.
- The rash of AE may mimic several medical conditions, such as necrolytic migratory erythema in glucagonoma syndrome especially in the setting of elevated glucagon.
- However, elevated glucagon is common in malnutrition as seen in our patient.
- **Differential diagnosis** of the typical erythematous scaly rash includes seborrheic dermatitis, psoriasis, pellagra, and impetigo.
- Medications associated with Zinc deficiency are also should be excluded as a cause such as PPIs, Loop diuretics, some ACEi and CCBs.
- Treatment of acquired AE involves lifelong zinc supplementation and clinical improvement can occasionally be promptly noticeable.

Conclusions

- Timely diagnosis and appropriate management are crucial as untreated AE can be life-threatening.
- Prompt clinical response is seen with zinc supplementation in AE.
- While zinc deficiency is more common in developing countries, it should be considered as a differential in patients with malnutrition and concurrent micronutrient deficiencies.