

BACKGROUND

- Zinc deficiency is commonly seen in developing regions of the world attributed to malnutrition.
- Acquired deficiency in developed countries are a combination of nutritional and chronic diseases/infections.
- Acrodermatitis enteropathica (AE) is a hereditary form, but acquired deficiencies can manifest with a similar clinical presentation.
- Untreated, zinc deficiency can lead to many complications related to skin, bone, and growth abnormalities.

Case Summary

- A 7-month-old unimmunized female of Amish background was admitted to the pediatric unit for 2-month history of worsening rash, fever, oral thrush, and diarrhea. The rash started on the face and thickened and progressed to cover part of the torso, genitals, buttocks and legs.
- She was prescribed topical corticosteroids as well as two full courses of oral clindamycin without relief.

Physical Examination

- Well demarcated erythematous confluent patches with overlying thick scales were present on the face, neck, lower torso, genitals, thighs.
- Oral thrush with thick white scales was present on the tongue and hard palate.

Figure 1. Anogenital and periorificial rash on initial presentation.



Case Summary contd.

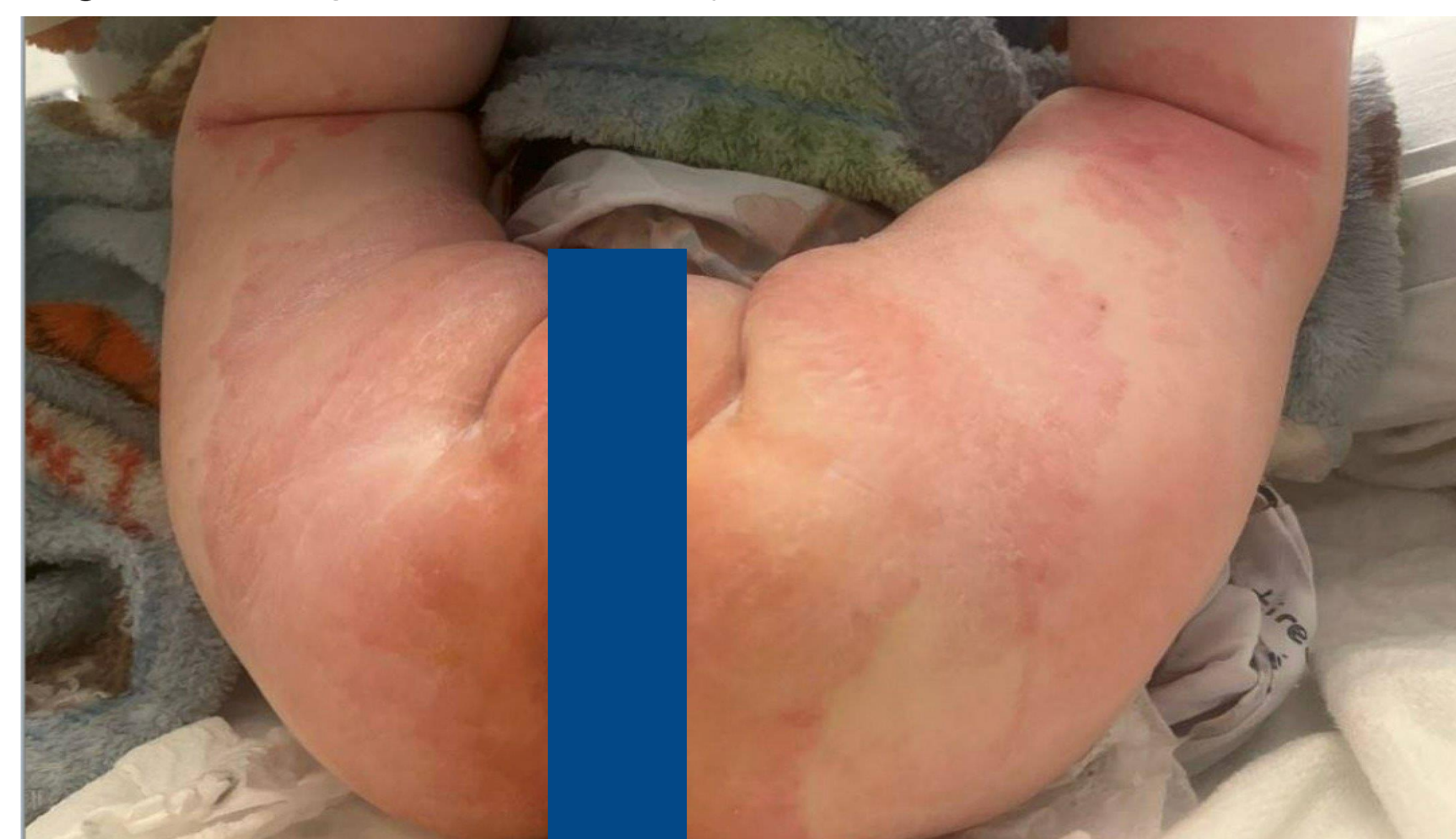
Work-up

- A full sepsis work up was pursued without a focus.
- CBC: thrombocytosis.
- CMP: Decreased alkaline phosphatase level
- Blood and urine cultures: Negative
- Skin lesion culture: *Candida albicans*, *Staphylococcus haemolyticus*, *Enterococcus faecalis*.
- Periorbital lesions: HSV-1 positive
- Respiratory panel: Parainfluenza type 1.

Management

- The patient received intravenous vancomycin, acyclovir, and cefazolin. IV fluconazole was added for mucocutaneous candidiasis.
- No clinically significant improvement was seen in the rash after 5 days of IV anti-microbials.
- Patient was noted to have watery stool and diarrhea throughout her admission.
- Serum zinc levels were ordered: revealed severe deficiency at 28 ug/dL
- Dermatology was consulted, and patient was clinically diagnosed with acrodermatitis enteropathica.

Figure 2. Anogenital rash on day 7 of admission.



- Patient was started on high-dose oral zinc sulfate.
- Rash improved significantly with antimicrobial therapy and zinc supplementation, and patient was discharged after one week of admission.

Discussion

- Acrodermatitis enteropathica is a hereditary form of zinc deficiency characterized by impaired zinc absorption in the gut due to a mutation of the SLC39A4 gene.²
- Clinical manifestations include periorificial and anogenital rash, diarrhea, and alopecia.
- Decreased levels of zinc-dependent enzymes such as alkaline phosphatase may be seen.²
- Differential includes acquired zinc deficiency due to dietary lack or low zinc levels in human milk. The patient was nearly exclusively breastfed at 7 months with minimal introduction to solids, which may have exacerbated her low zinc levels.³
- Return of persistent rash with discontinuation of zinc supplementation may support diagnosis of AE. Alternatively, sequencing of SLC39A4 gene may be considered.
- In infants and children presenting with perioral and anogenital rash, especially with minimal improvement on antimicrobial treatment, zinc deficiency must be considered as a potential cause. Evaluation of the patient should assess nutritional status, possible causes of malabsorption, as well as a hereditary etiology.
- Untreated zinc deficiency may have severe consequences. However, rapid improvement of cutaneous lesions is expected with prompt zinc supplementation.
- Our patient will be referred to genetics for testing for hereditary causes of zinc deficiency.

CONCLUSION

- Zinc deficiency, although rare, must be considered in infants and children presenting with periorificial and anogenital rash, especially if poor immune function is suspected. Assessment of dietary intake and nutrition is essential.

References

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