

BRIEF ARTICLE

Acrodermatitis Enteropathica Following a Pull-Through Procedure

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ABSTRACT

Introduction: Acrodermatitis Enteropathica is a rare condition traditionally expressed by perioral or acral dermatitis, diarrhea, and alopecia. Acrodermatitis Enteropathica can be inherited or acquired, with the acquired form linked to zinc deficiency. The pull-through procedure is a method used to treat Hirschsprung's disease, a condition in which the large intestine is missing nerve cells that make it difficult to pass stool, which can lead to obstruction and absorption issues. Complications of pull-through procedures have been associated with bowel obstruction and malabsorption of essential nutrients, including zinc.

Case Report: We present a 16-month-old male who developed perioral dermatitis, well-demarcated erythematous plaques, and alopecia after a pull-through procedure related to Hirschsprung's disease.

Conclusion: Acrodermatitis Enteropathica can present with various cutaneous lesions, and dermatologists should be familiar with the differing morphologies. In this paper, we encourage dermatologists to consider Acrodermatitis Enteropathica as a differential diagnosis for patients who develop cutaneous lesions following pull-through procedures.

INTRODUCTION

Acrodermatitis Enteropathica (AE) is a rare metabolic disorder that is linked to zinc deficiency.¹ AE is characterized by the presence of dermatitis (perioral, perianal, and acral), alopecia, and diarrhea.¹ The etiology of AE can be acquired or inherited. The acquired form of AE can result from a parent's inability to release zinc in breast milk, complications from certain abdominal surgeries, or from a lack of zinc in intravenous nutritional programs.² The hereditary form of the condition results from a genetic mutation in the SLC39A4 gene, a protein involved in zinc/iron-regulated

transportation and absorption.^{1,3} Zinc supplementation is the mainstay treatment to alleviate symptoms and improve health outcomes in patients afflicted by AE.

Hirschsprung's disease following pull-through procedures has been linked to cutaneous lesions with similar presentation to those found in zinc deficiency^{4,5}. Hirschsprung's disease is a congenital disorder in which there is abnormal development of the nerves supplying the large intestines in utero, disrupting the movement of stool in the digestive system. This can eventually lead to intestinal ischemia and malabsorption of nutrients, including zinc. The pull-through procedure

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involves the removal of abnormal bowel sections and pulling the remaining healthy segments through to normal bowel, connecting to the anus. In this case report, we explore the link between pull-through procedures associated with Hirschsprung's disease and cutaneous lesions related to zinc deficiency.

CASE REPORT

Our patient is a 16-month-old male with a history of Hirschsprung's disease status post a pull-through procedure. He presented to the clinic with well demarcated erythematous plaques in the perianal area, an erythematous crust to the perioral region, and erythema with silvery scale to their right second digit (**Figure 1**). According to the mother, his symptoms began shortly after the pull-through procedure was performed. The patient's symptoms previously failed to improve with over-the-counter diaper rash creams and miconazole cream. Our differential diagnoses included diaper dermatitis, histiocytosis, seborrheic dermatitis, and AE.

Fluticasone ointment was prescribed for the erythematous rash to the perianal area. A 3mm punch biopsy was performed and pathology revealed spongiotic dermatitis, chronic type with a PAS stain negative for fungal organisms. Additionally, the patient's zinc serum was 425ug/dl, below the lower limit of normal. We made sure that the lab used zinc-free tubes to not interfere with results. The patient later presented with increased hair loss and erythematous, greasy scales on his back, most likely seborrheic dermatitis.

Based on clinical characteristics and laboratory results, the presentation is suggestive of AE. The patient was started on

20mg of liquid zinc supplement daily (2mg/kg). The lesions began to subside within the first few weeks of treatment, and after two months they had completely resolved.

DISCUSSION

Zinc is an essential nutrient that is involved in cell division and growth, immune system function, and wound healing. While extreme zinc deficiency is rare in developed nations, it is the fifth leading cause of loss of healthy life years in developing countries.⁶ AE is a severe zinc-deficient state that can be expressed through a variety of skin lesions. The dermatological symptoms and histological characteristics of AE can also mimic several other nutrient deficient conditions, such as pellagra.⁷ Additionally, it is documented that patients who have undergone pull-through procedures related to Hirschsprung's disease have been shown to manifest with varying cutaneous lesions. One child after surgery at 4 months old presented later with moist, easily irritated, pustular lesions on scalp, hands, and feet with low zinc levels.⁴ In another case, a child with Hirschsprung's disease developed perianal pseudo-verrucous papules and nodules (PPPN) after their pull-through surgery.⁵ While clinical presentation of PPPN differs from AE, it is important to acknowledge that Hirschsprung's disease (and or pull-through procedures) is associated with several cutaneous lesions with differing morphologies.

In our case, AE was suspected due to the patient exhibiting two of the three classical physical manifestations of the condition; perioral and perianal dermatitis and alopecia, and a biopsy ruling out our remaining differential diagnoses (diaper dermatitis, histiocytosis, and seborrheic dermatitis). The



Figure 1. Initial presentation of lesions

diagnosis was confirmed by low serum zinc and the significant improvement of the patient's symptoms following the initiation of zinc supplementation. It is imperative to recognize the various disease conditions and surgical procedures that place patients at an increased risk for zinc deficiency. AE should be considered as a differential diagnosis for patients who develop cutaneous lesions with a history of Hirschsprung's disease status post pull-through procedures.

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References:

1. Nistor N, Ciontu L, Frasinariu OE, Lupu VV, Ignat A, Streanga V. Acrodermatitis Enteropathica: A Case Report. *Medicine (Baltimore)*. 2016;95(20):e3553. doi:10.1097/MD.0000000000003553
2. Acrodermatitis Enteropathica - Symptoms, Causes, Treatment | NORD. Accessed December 27, 2023. <https://rarediseases.org/rare-diseases/acrodermatitis-enteropathica/>
3. George AA, Mishra AK, Sahu KK, Sargent J. Acquired Acrodermatitis Enteropathica. *Am J Med*. 2021;134(1):e2-e3. doi:10.1016/j.amjmed.2020.09.011
4. Lien CH, Lin YL, Li ST, Lin CY. Skin lesion in a patient with Hirschsprung's disease complicated with short bowel syndrome: what is your diagnosis? *Case Rep*. 2012;2012:bcr0320126078. doi:10.1136/bcr-03-2012-6078
5. Yap FH, Thom G. Perianal pseudoverrucous papules and nodules in Hirschsprung's disease: Rapid resolution with oral loperamide. *Pediatr Dermatol*. 2017;34(6):e343-e344. doi:10.1111/pde.13275
6. Rabinovich D, Smadi Y. Zinc. In: *StatPearls*. StatPearls Publishing; 2023. Accessed December

- 27, 2023.
<http://www.ncbi.nlm.nih.gov/books/NBK547698/>
7. Giraldo-Villa A, Guzmán-Luna C. Acquired acrodermatitis enteropathica after abdominal surgery: Case report. *Case Rep.* 2019;5:147-154. doi:10.15446/cr.v5n2.78783