Acquired Acrodermatitis Enteropathica

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History of Present Illness: The patient presents with a desquamating mucocutaneous rash for one week. The rash began as tender, purple lesions which evolved into flaccid blisters of the abdomen, groin, extremities and palms. She admits to fatigue, mouth pain and skin tenderness. She denies dysphagia, hematuria, hematochezia, dysuria, painful defecation.

Medical History/Surgical History: Liver cirrhosis, nonalcoholic steatohepatitis, hepatic encephalopathy, iron deficiency anemia, morbid obesity, vitamin B12 deficiency, vitamin D deficiency, biliopancreatic diversion with duodenal switch, cholecystectomy

Family History: Hypertension

Medications: ciprofloxacin, famotidine, furosemide, gabapentin, hydroxyzine, lactulose, melatonin, ranitidine, rifaximin, spironolactone, thiamine, tramadol, vitamin A, vitamin D

Previous Treatments: Zinc sulfate 220 mg oral daily, zinc 61 mg intravenous daily, multitrace®-5 concentrate 6mL IV (manganese, chromium, selenium, zinc, copper), multivitamin 5mL IV daily (biotin, folic acid, vitamins A, B1, B3, B5, B-12, C, D, E, K)

Current Treatment: B-complex with vitamin C, Theragran® multivitamin

Physical Examination: Moist erosions of the lateral commissures. Desquamation of the bilateral palms. Moist denuded erosions of the abdomen and extremities. Flaccid bullae on the lower extremities and dorsal feet.

Laboratory Data: Zinc 20 ug/dL (60-120), tryptophan 3 umol/L (40-91), copper 20.7 ug/dL (80-155), vitamin B6 25 nmol/L (20-125), methylmalonic acid 0.19 umol/L (0.0-0.40), albumin 2.2 g/dL (3.4-5.4). CMP, ANA, hep panel, TSH WNL. Aerobic, anaerobic, viral cultures of abdomen negative.

Biopsy: *Health Network Laboratories* (S19-31696, 06/23/19) Left leg: "Upper epidermal keratinocytic vacuolated change with an overlying zone of confluent parakeratosis and underlying clefting and mild spongiosis. PASD negative."

Mayo Medical Laboratories (DR-19-16862, 06/23/19) Left leg, direct immunofluorescence: "Weak discontinuous granular C3 deposition along the basement membrane zone. Patchy dermal fibrinogen deposition. IgG, IgM, IgA negative."

Figure 1: Erosions of the lateral commissures of the mouth with hemorrhagic crust.

Figure 2: Denuded skin with underlying moist erosions with pink bases in the gluteal cleft and medial buttocks.

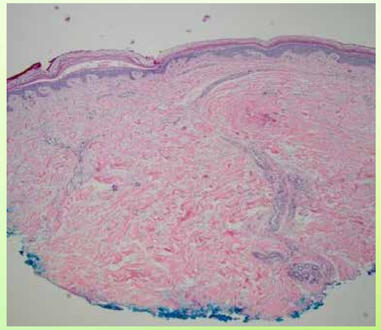

Figure 3: H&E, Left thigh (10x): Intraepidermal clefting with underlying spongiosis and mild superficial perivascular inflammation.

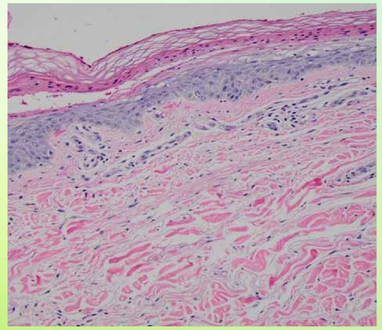

Figure 4: H&E, Left thigh (40x): Upper epidermal keratinocytic vacuolated change with an overlying zone of confluent parakeratosis and underlying clefting and mild spongiosis.

REFERENCES

[1]Freitas J, Bliven P, Case R. Combined zinc and vitamin B6 deficiency in a patient with diffuse red rash and angular cheilitis 6 years after Roux-en-Y gastric bypass. *BMJ Case Reports* CP 2019;12.

[2]Macdonald J, Connolly S, DiCaudo D. Think Zinc Deficiency: Acquired Acrodermatitis Enteropathica Due to Poor Diet and Common Medications. *Arch Dermatol.* 2012;148(8):961–963.

[3]Kurt BÖ et al. Acquired acrodermatitis enteropathica secondary to sleeve gastrectomy. *Indian J Dermatol Venereol Leprol.* 2019;85:220-3.

Diagnosis: Acquired Acrodermatitis Enteropathica

Acrodermatitis enteropathica (AAE) is a rare disorder within the family of necrolytic erythema or nutritional deficiency dermatoses. It is caused by decreased dietary zinc intake, malabsorption, or increased urinary excretion. In adults, acquired acrodermatitis enteropathica may be caused by anorexia nervosa, alcoholism, intestinal malabsorption, diets high in mineral binding phytate, or certain medications. Hypertensive medications such as thiazides, loop diuretics, angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers increase urinary excretion of zinc.

AAE is clinically characterized as a papulosquamous dermatitis with secondary erosions or ulcerations involving periorificial, intertriginous, and acral skin. Associated signs and symptoms include alopecia, nail dystrophy, intraoral erosions, ocular disease, diarrhea, hypogeusia, hyposmia, and lethargy. However, necrolytic erythema is a non-specific finding common to many nutritional deficiencies.

Diagnostic studies include evaluation of serum zinc levels and skin biopsy. The normal serum zinc level is 66 to 110 µg/dL. In addition, serum alkaline phosphatase can be checked to help diagnose a zinc deficiency. Alkaline phosphatase is a zinc-dependent enzyme and thus levels are low in zinc deficiency. Zinc is albumin-bound; therefore, you must account for albumin levels in conjunction with your serum zinc levels. Histopathologic features include focal parakeratosis, hypogranulosis, keratinocyte dysmaturation, and epidermal pallor.

Treatment of AAE includes dietary supplementation with elemental zinc 0.5-1.0 mg/kg/day. A 220-mg zinc sulfate tablet contains 50 mg of elemental zinc. Cutaneous manifestations improve within days of initiating treatment, and it is recommended to continue until serum levels normalize. The American Society for Metabolic and Bariatric Surgery Nutrition guidelines after Roux-en-Y gastric bypass and biliopancreatic diversion recommend annual screening of serum levels of zinc, copper, calcium, folate, iron, vitamins A, B1, B12, D, E, and K.

Rising obesity and the increased prevalence of bariatric surgeries may result in an increased incidence of nutritional deficiency dermatoses in patients after bariatric surgery, especially malabsorptive procedures like biliopancreatic diversion or Roux-en-Y gastric bypass. Despite biliopancreatic diversion being the most effective bariatric procedure for weight loss, it represents less than 2% of bariatric surgeries worldwide. Its diminished use is due in part to excessive malabsorption leading to protein and vitamin deficiencies.