# Desquamative erythematous rash in a young woman

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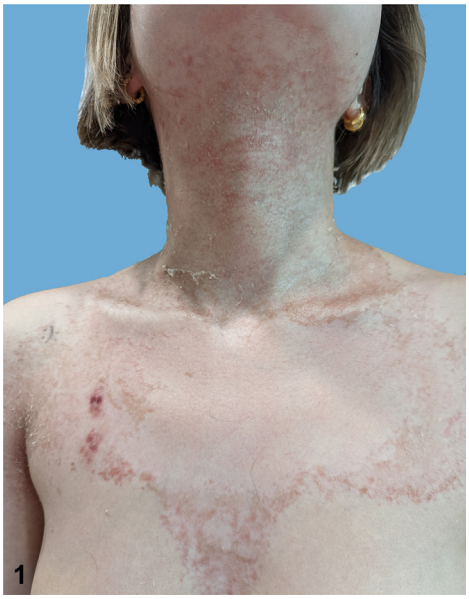
***Key words:*** biotin; biotin deficiency; granular parakeratosis; nutrition; nutritional deficiency; vitamin deficiency; vitamin H.



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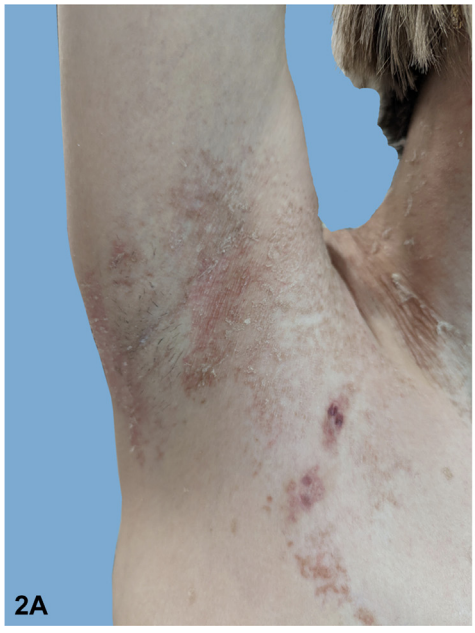

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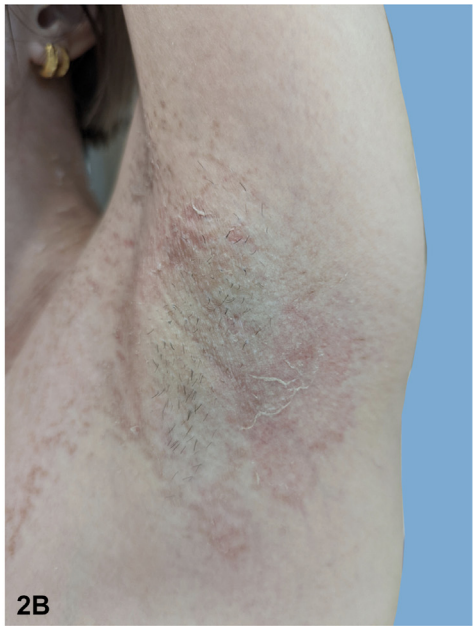

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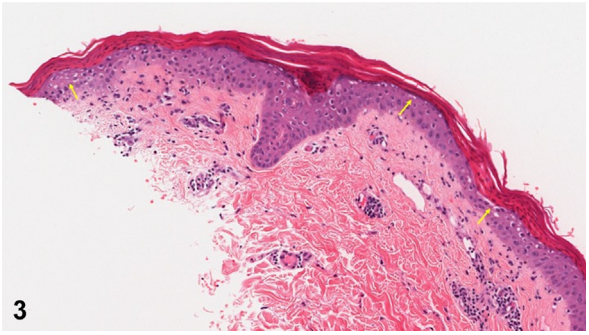


A 22-year-old woman presented with a worsening nonpruritic rash on the anterior aspect of her chest and underarms for the past 6 months. The rash was associated with dryness, desquamation, and a burning sensation. She had no preceding new contactants or medications. In addition, she had discontinued all contactants upon the onset of her rash; however, her condition continued to progress despite the cessation of contactants for more than a month. Her rashes also persisted despite continual use of topical corticosteroids and topical tacrolimus. She reported going on a crash diet one year ago for weight loss and lost a total of 6 kg in the first 3 months. Currently, she only consumes 500 to 800 calories a day and currently weighs 38 kg with a body mass index of 16.78 kg/m$^2$. She denies any bulimic symptoms, recent stressors, or mood problems. Her medical history is significant for asthma, allergic rhinitis, and a previous anxiety disorder.

Examination revealed sharply demarcated, erythematous patches, with superficial erosions and desquamative scales, on her anterior aspect of the neck, extending to both clavicles, upper portion of the chest, axilla, sternum, and left inframammary region (Figs 1 and 2). The flexural rashes were associated with hyperesthesia and hyperalgesia. The rest of the systemic review and examination were unremarkable.

A punch biopsy of the upper portion of the right side of the chest showed granular parakeratosis and mild epidermal dysmaturation with upper epidermal vacuolation and pallor (Fig 3).

**Question 1: What is the most appropriate next step of management?**

**A.** Skin fungal microscopy and culture

**B.** Myositis-specific antibodies

**C.** Serum biotin, niacin, and zinc levels

**D.** Patch testing

**E.** Treat with oral steroids

**Answers:**

**A.** Incorrect — the overall suspicion of cutaneous fungal infection was not high in this patient, especially with the morphology of superficial erosions associated with hyperesthesia and the absence of fungal elements on the skin biopsy histology.

**B.** Incorrect — Some may mistake the neck and chest rashes for the scarf/shawl sign in dermatomyositis, but the rash was not photo-distributed and it does not show violaceous erythema or poikiloderma. There were no characteristic associated features, such as heliotrope rash, Gottron's papules, periungual erythema, or myopathy.

**C.** Correct — The key to the correct diagnosis, in this case, is the combination of the morphology of an eruption of erythematous patches and plaques on her neck and flexures with superficial erosions and desquamative scales, coupled with the histologic

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findings of granular parakeratosis with upper epidermal pallor and history of eating disorder — suggesting an underlying nutritional disorder.

**D.** Incorrect — There was no history of any new contactants in this patient, thus patch testing would likely be unyielding. Furthermore, this would not address the underlying condition of nutritional disorder.

**E.** Incorrect — Although oral steroids may temporarily improve the dermatitis of a nutritional deficiency, it would not address the underlying cause of her dermatitis, thus it is not the most appropriate therapy.

**Question 2: What is the most likely diagnosis?**

**A.** Zinc deficiency

**B.** Niacin deficiency

**C.** Biotin deficiency

**D.** Vitamin A deficiency

**E.** Protein deficiency

**Answers:**

**A.** Incorrect — Zinc deficiency leading to acrodermatitis enteropathica may present similarly with sharply demarcated erythematous, dry, scaly patches with superficial erosions. However, she did not have the typical distribution of rashes involving the periorificial, anogenital, and acral regions.[1] There was also an absence of alopecia, nail changes, and diarrhea. Eventually, her serum zinc levels were found to be 754 ug/L (normal: 724-1244 ug/L).

**B.** Incorrect — Niacin deficiency leading to pellagra may present with rashes on the neck and upper portion of the chest resembling a "Casal necklace," a clinical sign comprising of a well-demarcated band of erythematous and hyperpigmented scaly rash around the neck.[2] However, her flexural rashes were not photo-distributed. She had no other systemic symptoms besides deliberate weight loss. However, her niacin levels returned normal at 2.45 ug/mL (normal: 0.5-8.45ug/mL).

**C.** Correct — Biotin deficiency was suspected as biotin is a cofactor for zinc homeostasis[3] in the skin. Mucocutaneous manifestations of biotin deficiency usually present 3 to 6 months after the onset of deficiency. They include acrodermatitis enteropathica-like rashes (often periorificial), erosions on flexural folds, alopecia, blepharoconjunctivitis, onychoschizia, and hyperesthesia.[4] A low-serum biotin level was found in our patient at 0.2 ng/mL (normal ≥0.3 ng/mL). Although she did not present with all the mucocutaneous features of biotin deficiency, the replacement of biotin resulted in significant improvement and eventual resolution of her rashes within a few weeks. There has been no recurrence while she was on biotin supplementation. We postulate that the rashes may favor flexural location due to bending movements and friction, which cause the breakdown of the epidermis composed of degenerating keratinocytes due to the underlying nutritional deficiency. The patient was eventually prescribed high-dose oral biotin supplementation (10,000 mcg twice daily) in addition to multivitamins with the improvement of her rashes within a few days, which subsequently resolved in 3 weeks. She remains free of recurrence over the next one year after receiving biotin supplementation. In addition, she was also referred to a psychiatrist and was diagnosed with anorexia nervosa.

**D.** Incorrect — Patients with vitamin A deficiency may manifest as phrynoderma, which are clusters of follicular papules with central keratotic plugs. Histologic findings include keratinous plugs within follicles, hyperkeratosis, and sebaceous gland atrophy. Our patient did not have such skin or histologic findings, and her vitamin A levels were 0.46 mg/L (normal: 0.26-0.72 mg/L).

**E.** Incorrect — Kwashiorkor is an acute form of protein-energy malnutrition. Although patients with kwashiorkor can develop erythematous rashes with erosions and desquamation resembling "flaky paint," characteristic findings are peripheral edema or anasarca in association with hypoalbuminemia. This patient did not have any peripheral edema, and her serum albumin levels were normal at 44 g/L (normal: 38-48 g/L).

**Question 3: Which of these manifestations are not associated with biotin deficiency?**

**A.** Alopecia

**B.** Seizures

**C.** Hearing loss

**D.** Diarrhea

**E.** Metabolic acidosis

**Answers:**

**A.** Incorrect − Alopecia is a common cutaneous symptom of biotin deficiency.[4] Other common cutaneous symptoms include periorificial dermatitis and flexural rashes.

**B.** Incorrect − The most common neurologic feature of patients with profound biotin deficiency is seizures and hypotonia. The seizures are usually myoclonic but may be grand mal and focal.[5]

**C.** Incorrect − Sensorineural hearing loss has been described in untreated cases of biotin deficiency. Unfortunately, the hearing loss associated with biotin deficiency usually does not resolve or improve but remains static with biotin replacement. These cases are usually treated with the use of hearing aids or implants.

**D.** Correct − Diarrhea is not a manifestation of biotin deficiency. The presence of diarrhea is seen more often in cases of niacin deficiency, in which patients present with a constellation of symptoms including diarrhea, dermatitis, and dementia.

**E.** Incorrect − Most untreated patients with biotin deficiency will develop metabolic acidosis, contributed by the presence of ketoacidosis, lactic acidosis, and/or hyperammonemia.[5]

**Conflicts of interest**

None disclosed.

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