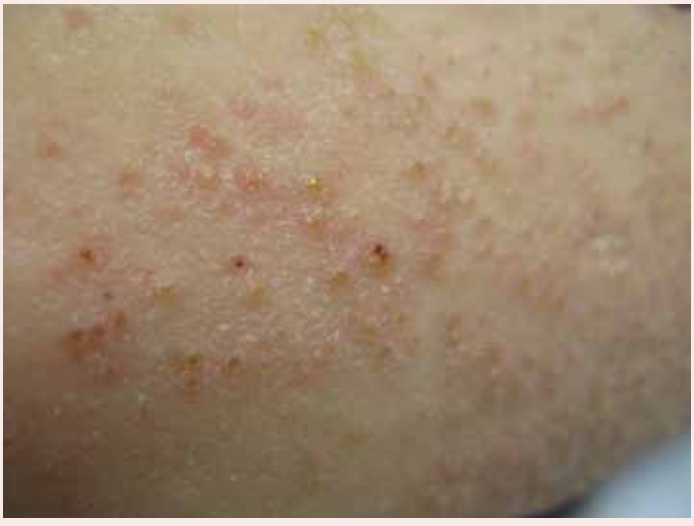
Dry Scaly Rash

What's the diagnosis?



A 22-year-old man with a history of severe mental retardation, a questionable diagnosis of cystic fibrosis, chronic abdominal pain, and a recent diagnosis of primary sclerosing cholangitis was transferred from an outside hospital for worsening abdominal pain, lethargy, anasarca, anorexia, and constipation. An abdominal paracentesis and computed tomography scan of the abdomen were both negative for any acute processes. Blood cultures and paracentesis fluid cultures were negative. The dermatology section was consulted for a dry scaly rash of unknown duration. The eruption had not been treated, and the patient's dermatologic history was otherwise unremarkable. On physical examination, the patient had a generalized follicular, erythematous, and scaly papular eruption with some areas of yellow-colored hyperkeratotic papules and plaques as well as scattered hemorrhagic crusts on the extensor surface of the right arm. He had normal oral mucosa and nails. The team was unable to gain consent for biopsy, but a blood test was obtained.

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The authors report no conflict of interest.
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The Diagnosis: Phrynoderma

Phrynoderma, meaning "toad skin," was first described by Nicholls[1] in 1933 as a condition due to vitamin A deficiency and characterized by follicular hyperkeratosis. Since then, there has been debate in the literature over the etiology of phrynoderma, as studies have demonstrated different causes for this condition,[2-5] which suggests that it is prudent to check several markers of nutritional status including vitamin A, vitamin B, vitamin E, and essential fatty acid studies.

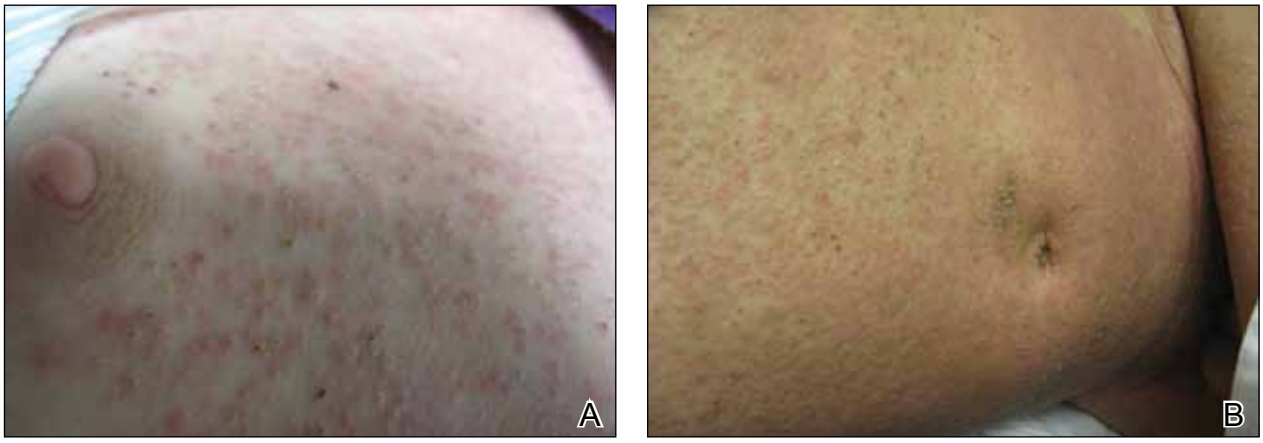
Phrynoderma is most commonly seen in South and East Asia with relatively rare occurrences in the United States. Although it characteristically occurs in children and adolescents aged 5 to 15 years, almost all of the cases reported in the United States have occurred in adults.[6,7] A clear gender predilection has not been established.[7]

Phrynoderma tends to favor the extensor surfaces. In particular, the elbows, thighs, and buttocks are the most commonly affected locations. The face is typically the last site to become involved. On physical examination of a patient with phrynoderma, there are various-sized, discrete, red-brown to flesh-colored papules with a conical keratotic follicular plug (Figure).[3] The eruption is often asymptomatic, but mild pruritus may be reported.[7] Additional signs of vitamin A deficiency may be present, including night blindness, hyperpigmentation, or xerosis. Patients with phrynoderma also may have evidence of other nutritional deficiencies, such as angular stomatitis, cheilitis, or glossitis. The differential diagnosis for a patient with this type of papular eruption may include pityriasis rubra pilaris, keratosis pilaris, lichen spinulosus, crusted scabies, and follicular lichen planus.[7] History and clinical presentation can generally help to distinguish these diagnoses.

Histopathology typically shows moderate hyperkeratosis with distension of the upper portion of the follicle by large keratotic plugs. Sebaceous glands are greatly reduced in size and may exhibit epithelial atrophy.[8,9]

A reduced serum vitamin A level can confirm the diagnosis of phrynoderma. Our patient's laboratory results revealed a vitamin A value of 26 μg/100 dL (reference range, 38–106 μg/100 dL), consistent with his clinical presentation of phrynoderma. Other markers of nutritional status, including vitamins D, E, and K_1, were all within reference range. Treatment consists of replacing the nutritional deficit. High doses of vitamin A, such as 200,000 to 500,000 IU daily for 3 to 5 days or daily doses of 2000 to 50,000 IU, can lead to resolution of the rash in 1 to 4 months.[8,9] Our patient was started on 10,000 IU of oral vitamin A daily at discharge but unfortunately died a few months later.

Vitamin A has been shown to promote maturation and differentiation of the basal keratinocytes as they move up through the epidermis, which may in part explain why this treatment is effective.[10] It is important to be aware of this condition when seeing a patient with follicular hyperkeratosis because even patients in developed countries with malnutrition due to diet or gastrointestinal disease may develop phrynoderma.



Generalized follicular, erythematous, and scaly papular eruption on the chest (A) and abdomen (B).

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