

# Keratosi s Circumscripta Revisited: A Case Report and Review of the Literature

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## GOAL

To understand keratosi s circumscripta to better manage patients with the condition

## OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical presentation of keratosi s circumscripta.
2. Discuss the disorders often compared to keratosi s circumscripta.
3. Identify treatment options for keratosi s circumscripta.

**CME** Test on page 378.

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*Keratosi s circumscripta, also known as psoriasis circumscripta with palmoplantar keratosis, is a rarely reported condition that manifests as well-circumscribed lesions consisting of grouped folliculocentric papules on the elbows, knees, neck, sacrum, posterior axillary folds, and hips. This condition typically begins in childhood and has*

*been shown to improve, but not resolve, with keratolytic therapies. Some debate exists concerning the terminology used to identify this condition. Our report contributes to this ongoing dialogue by presenting a case that supports recognizing keratosi s circumscripta as a unique clinical entity. We describe the diagnosis and treatment of a boy with keratosi s circumscripta. We also present a review of the literature pertaining to this condition and an overview of the controversy surrounding its terminology.*

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## Case Report

An 8-year-old African American boy presented in the summer of 2005 with a 5-year history of a rash on his elbows, knees, and hips. The patient's lesions



Keratosis circumscripta with symmetric areas of sharply demarcated follicular hyperkeratosis and scale in a background of thickened violaceous plaques on the extensor surfaces of the elbows and knees.

initially developed at 3 years of age and were described by his pediatrician as small 1- to 2-mm hyperkeratotic perifollicular papules on the elbows, knees, and hips that were hardly noticeable. From 3 to 6 years of age, the involved areas slowly developed into diffuse violaceous plaques that became thickened and markedly more demarcated from the surrounding unaffected skin. For the 2 to 3 years prior to presentation to our clinic, the child's lesions remained stable and did not wax or wane independently. The patient never developed new lesions on other parts of his body. The lesions did not improve or resolve with sun exposure and remained unresponsive to all therapies initiated by the child's pediatrician, including various steroid creams and emollients such as hydrocortisone 1%, hydrocortisone 2.5%, triamcinolone acetonide 0.1%, Aquaphor<sup>®</sup>, Eucerin<sup>®</sup>, and Cetaphil<sup>®</sup>.

The boy was born in the United States and was the third generation of an American family that emigrated from Africa. The child's parents denied knowing where in Africa their family had resided. The child had no known relatives who were similarly affected.

Results of a physical examination revealed symmetric areas of sharply demarcated follicular hyperkeratosis and scale in a background of thickened violaceous plaques on the extensor surfaces of the patient's elbows and knees (Figure) as well as symmetric lesions on each hip. The lesions were not erythematous, showed no scale between the follicles, and were not micaceous in nature. The patient had no skin thickening of the palms and soles or scaling of the scalp or face. The patient also did not have any lesions on his neck, buttocks, abdomen, shoulders, popliteal fossae, or extensor surfaces of his arms.

Four weeks after initiation of urea cream 40% applied twice daily to the affected areas, the patient's lesions were softer, smoother, and considerably less hyperkeratotic, with resolution of the perifollicular

papules. There was no change in color and the violaceous plaques still were sharply demarcated. Maintenance therapy with urea cream 40% provided sustained results with which the patient and his parents were extremely satisfied.

### Comment

Originally described by Shrank<sup>1</sup> in 1966, keratosis circumscripta typically is associated with individuals of African descent, specifically descendants of the Yoruba tribe of Nigeria. Shrank<sup>1</sup> identified lesions in 11 patients, all between the ages of 3 and 17 years and of the Yoruba tribe of Nigeria. The lesions were described as "sharply defined areas of diffuse and follicular hyperkeratosis on the hands, feet, elbows, knees, and trunk." The extensor surfaces of the elbows and knees always were affected, as was the trunk in the form of circular disks of follicular hyperkeratosis on each hip. The backs of the hands and dorsa of the feet occasionally were affected, but the palms, soles, and nails rarely were affected. Biopsy results demonstrated follicular plugging and moderate hyperkeratosis, with the lower layers of the epidermis and dermis unaffected. The lesions developed in 2 to 3 weeks between the ages of 3 and 5 years, and they did not spread any further. Notably, Shrank<sup>1</sup> reported no adult cases.

Shrank<sup>1</sup> initially thought that the disorder was an acquired dermatosis. He specifically suspected hypervitaminosis A because the Yoruba tribe was known to cook much of their food in red palm oil, a substance rich in vitamin A. He discounted this diagnosis after determining that the patients' serum levels of  $\beta$ -carotene and vitamin A were within reference range. For the same reasons, he discounted a deficiency of vitamin B complex. The author also suspected juvenile pityriasis rubra pilaris (PRP). Although he noted that the disorder shared some similarities with juvenile PRP, he reported that the children failed to improve with topical steroid and oral vitamin A treatments. Also, his patients did not exhibit diffuse erythema or scaling of the scalp and face, findings that are associated with juvenile PRP. Thus, Shrank<sup>1</sup> concluded that sufficient differences existed between the conditions to merit recognizing the disorder as a separate entity.

Shrank<sup>1</sup> then considered that the condition could have represented a recessively inherited genodermatosis, noting that the children were all from the same tribe and that their mothers and fathers were not affected. However, he found no evidence of any affected siblings, a strong argument against the disorder being an inherited condition. He also considered 2 genodermatoses that closely resembled the clinical appearance of the cases that he followed.<sup>1</sup>

These disorders were mal de Meleda and a hereditary condition described by Greither.<sup>2</sup> Like the condition observed by Shrank,<sup>1</sup> both of these genodermatoses exhibit hyperkeratosis of the hands, feet, elbows, and knees; however, the trunk is never involved as it is in keratosis circumscripta. Additionally, in mal de Meleda, the palms, soles, and nails always are involved, and the disease progresses through life, with eventual involvement of the elbows and knees.<sup>3</sup> The genodermatosis described by Greither<sup>2</sup> was not consistent with what Shrank<sup>1</sup> observed because it demonstrated an autosomal dominant mode of inheritance. While maintaining that his cases could represent a genodermatosis inherited as an autosomal-recessive trait, Shrank<sup>1</sup> concluded that the disorder he observed had sufficient differences from these 2 genodermatoses for it to be regarded as a separate entity, and he named the condition *keratosis circumscripta*.

In 1978, Soyinka and Laja<sup>4</sup> published a report that countered Shrank.<sup>1</sup> Rather than keratosis circumscripta, they proposed that Shrank's<sup>1</sup> patients suffered from an environmentally modified variant of psoriasis. Upon investigating Shrank's<sup>1</sup> original records, the authors reported that 4 of 11 biopsies were recorded as having had many characteristics that, in their opinion, reflected a variant of psoriasis. In particular, these characteristics included "destruction of basal layer with lymphocytic infiltrations," "elongation of the rete pegs," "pointed rete ridges," and "occasional intracellular oedema."<sup>4</sup> Interestingly, Soyinka and Laja<sup>4</sup> remarked that Shrank<sup>1</sup> never referred to these characteristics in his 1966 article and only described follicular plugging, moderate hyperkeratosis, and occasional parakeratosis. Based on their review of Shrank's<sup>1</sup> work, Soyinka and Laja<sup>4</sup> proposed renaming this condition *psoriasis circumscripta with palmoplantar keratosis*, a term that has since been used interchangeably with *keratosis circumscripta* despite the lack of evidence definitively linking the 2 conditions to one another.

In 1979, Verhagen<sup>5</sup> continued the dialogue on this disorder, responding to Soyinka and Laja's<sup>4</sup> conclusions and defending Shrank's<sup>1</sup> findings. Referring to his experiences in Kenya where he encountered about 10 cases similar to Shrank's<sup>1</sup> patients, Verhagen<sup>5</sup> reported that he found minimal similarities between keratosis circumscripta and psoriasis. His patients showed a type of hyperkeratosis "totally different" from that of psoriasis and exhibited "hardly any dermal abnormalities, notably in the papilla, or Munro's abscesses."<sup>5</sup> Moreover, like those patients described by Shrank,<sup>1</sup> Verhagen's<sup>5</sup> patients did not respond to corticosteroids. Also, Verhagen<sup>5</sup> noted

that his patients' lesions were stable over a period of years and never resembled psoriasis.

A contemporary text describes keratosis circumscripta as having folliculocentric papules grouped into well-circumscribed areas, predominately on the elbows, knees, neck, sacrum, posterior axillary folds, and hips.<sup>6</sup> Although similar to psoriasis, the histologic findings of keratosis circumscripta reportedly do not show neutrophilic microabscesses, and no evidence exists of patients with the disorder who eventually developed psoriasis. The text notes that lesions of keratosis circumscripta may improve with exfoliant/emollient preparations such as topical urea cream 40%, salicylic acid cream 2% in urea cream 20%, or glycolic acid preparations. Salicylic acid peels and laser treatments also may prove to be therapeutic.<sup>6</sup>

Kelly<sup>6</sup> noted that keratosis circumscripta clinically overlaps with juvenile PRP and lichen spinulosus. Although all 3 conditions have well-defined plaques of hyperkeratoses on the elbows and knees, the overall presentation and histology of keratosis circumscripta are sufficiently different from juvenile PRP and lichen spinulosus, warranting its classification as a separate clinical entity. The histologic findings in juvenile PRP are similar to those of keratosis circumscripta, with follicular plugging and moderate hyperkeratosis, and with the lower layers of the epidermis and dermis unaffected. However, as observed in our patient, keratosis circumscripta typically does not have the thickening of the skin of the palms and soles, which often is found in juvenile PRP. Also, our patient's lesions did not demonstrate any erythema, and he did not have scaling of the face or scalp as can be found in juvenile PRP. Additionally, juvenile PRP lacks the circular discs of follicular hyperkeratosis that were found on the hips of our patient, which are consistent with a diagnosis of keratosis circumscripta.<sup>7</sup> With respect to lichen spinulosus, our patient did not have plaques on the buttocks, abdomen, shoulders, popliteal fossae, or extensor surfaces of his arms as is usually seen with this disorder. His lesions enlarged relatively slowly, rather than the rapid fashion reported in patients with lichen spinulosus. Furthermore, our patient demonstrated no erythema and his plaques were much more sharply demarcated than those found in lichen spinulosus.

The controversy surrounding the naming of keratosis circumscripta has resulted in the pseudonym *psoriasis circumscripta with palmoplantar keratosis*, which implies that the disorder is a variant of psoriasis. However, our case report and review of the literature suggest that keratosis circumscripta

exhibits enough unique characteristics to warrant designating it as a separate clinical entity. The name *psoriasis circumscripta with palmoplantar keratosis* would not fit our patient's clinical presentation because he had no palmar or plantar lesions. The patient's well-circumscribed plaques had distinct follicular hyperkeratosis and scale, and the lesions were in a background of thickened violaceous plaques. However, there was no scale between the follicles, which is inconsistent with psoriasis. The patient's scale was not micaceous and thick, and there was a complete lack of erythema, both of which are common in psoriasis. The patient's lesions grew larger for approximately 3 years and then remained stable, unlike lesions in psoriasis, which often worsen throughout life. After 6 years of age, the patient's lesions did not worsen with time; they also did not lessen or resolve with sun exposure. Additionally, the lesions did not respond to topical steroid treatments. All of these findings are atypical for psoriasis, and the history, distribution, and characteristics of this patient's lesions resembled those originally described by Shrank.<sup>1</sup>

The term *keratosis circumscripta* offers a diagnostic clarity that cannot be accurately achieved by using the term interchangeably with *psoriasis circumscripta with palmoplantar keratosis*. In our patient, the term *psoriasis circumscripta with palmoplantar keratosis* is in fact a misnomer because the patient did not have any palmoplantar lesions and his lesions did not resemble psoriasis. Unlike *psoriasis circumscripta with palmoplantar keratosis*, the term *keratosis circumscripta* does not suggest to a physician that corticosteroid therapy is an appropriate treatment modality. Our patient did not respond to corticosteroid therapy, a course of intervention that would have likely improved his lesions had they been caused by a variant of psoriasis. Furthermore, *keratosis circumscripta* should be regarded as a distinct clinical entity because if the disorder is later determined to be a genodermatosis, a possibility suggested by Shrank,<sup>1</sup> its specific manifestations may facilitate our understanding of the genetic mechanisms particular to its expression.

## Comment

Our clinical findings suggest that our patient had *keratosis circumscripta*. This disorder has a distinctive symmetric, stable, nonprogressive, and sharply demarcated clinical presentation. It demonstrates resistance to treatments that would normally improve clinical entities such as psoriasiform lesions, juvenile PRP, and lichen spinulosus, all of which share some characteristics associated with this disorder. *Keratosis circumscripta* clinically exhibits folliculocentric papules grouped into well-circumscribed clusters on the elbows, knees, neck, sacrum, posterior axillary folds, and hips. Histologically, *keratosis circumscripta* shows follicular plugging and moderate hyperkeratosis, with the lower layers of the epidermis and dermis unaffected and showing no evidence of neutrophilic microabscesses. The condition appears to respond to keratinolytic therapies. Based on our clinical findings and review of the literature, we conclude that *keratosis circumscripta* is a unique clinical entity and that distinguishing it as such will facilitate our ability to understand and appropriately treat this rare and unique condition.

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