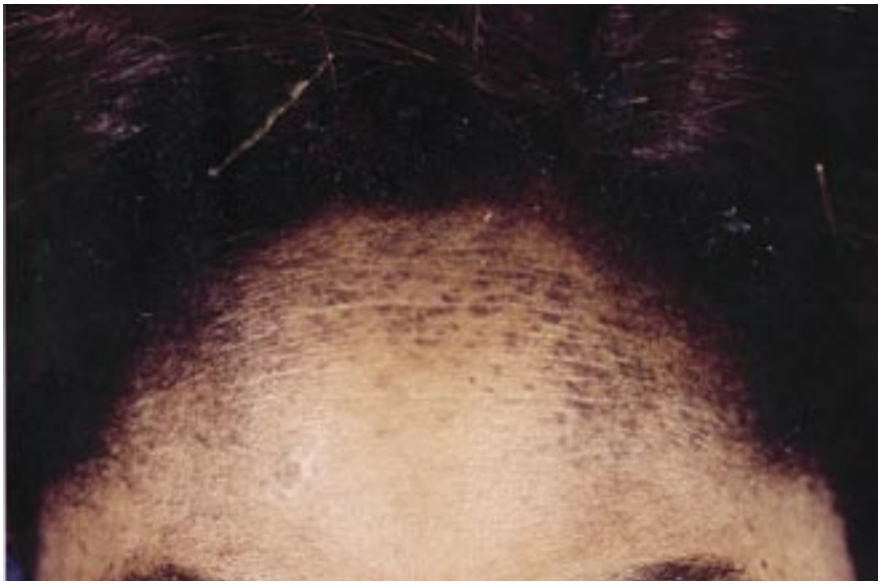
What Is Your Diagnosis?



A 33-year-old Caribbean woman presented with a 2-year history of an asymptomatic skin eruption that initially affected her back but slowly spread to involve her upper forehead and postauricular region. Her past medical history was unremarkable; there was no history of diabetes mellitus and no family history of a similar cutaneous eruption. Physical examination revealed a mildly obese woman with grayish brown, hyperkeratotic, 3- to 4-mm papules that coalesced to form diffuse large plaques with a reticulate peripheral pattern involving the upper back, upper forehead, and behind the ears. The scalp could not be examined because of a tight hair weave. Wood lamp examination revealed no accentuation or diminishment of pigmentation. A potassium hydroxide smear was negative for organisms. Physical examination also revealed acanthosis nigricans involving the neck and bilateral axillae.

PLEASE TURN TO PAGE 239 FOR DISCUSSION

Josephine Nguyen, MD; Christopher Norwood, MD; National Naval Medical Center, Bethesda, Maryland.
The authors report no conflict of interest.
The assertions or opinions contained herein are the private views of the authors and are not to be construed as official or as reflecting the views of the US Department of the Navy or the US Government.

The Diagnosis: Confluent and Reticulate Papillomatosis (Gougerot-Carteaud Syndrome)

Skin biopsy of the diffuse large plaques with a reticulate peripheral pattern (Figure 1) showed papillomatosis and hyperkeratosis (Figure 2). Periodic acid-Schiff stain was negative. The patient had normal blood chemistries and complete blood profile. Given the clinical presentation and typical, though nonspecific, histopathologic findings, a diagnosis of confluent and reticulate papillomatosis (CRP)(Gougerot-Carteaud syndrome) was made. The patient was treated with minocycline 200 mg/d, and on one-month follow-up, showed 95% clearance, with no effect on the velvety axillary and neck plaques, and complete resolution after 2 months.

CRP (Gougerot-Carteaud syndrome) is a rare condition that was first described in 1927.[1] The lesions usually begin after puberty, more commonly in females.[2] There have been 2 familial cases reported in the literature, but most cases are sporadic.[3,4] The eruption usually begins as brown reticulate papules with a fine scale that originate in the intermammary and interscapular regions and progressively spread to the breast and abdomen. There has been only one reported case of CRP localized to the cheeks,[5] but to the best of our knowledge, there have been no reports of involvement of the forehead.

The differential diagnosis of Gougerot-Carteaud syndrome includes acanthosis nigricans, which involves the intertriginous areas, such as the neck and axillae; tinea versicolor; pseudoacanthosis nigricans, which is associated with obesity and disappears with weight loss; Darier disease; and hereditary reticulate dyschromatoses.[6,7]

CRP remains an enigma. The pathophysiology of this disorder is still unknown. The 2 main theories suggest that there is an abnormal host response to *Pityrosporum orbiculare* colonization and a defect in keratinization. CRP has been associated with *P orbiculare* because the distribution of the eruption corresponds to the area usually colonized by the organism[8] and the documented improvement with antifungal therapy in a number of cases.[5,8] However, *Pityrosporum ovale* is not isolated in all cases and response to antimycotic therapy has been variable. A keratinization defect has been suggested because of the response to retinoid therapy.[9-12] Under electron microscopy, an increased number of transitional cells can be found between the stratum granulosum and stratum corneum, also indicating that CRP could be a keratinization disorder.[13] Jang et al[14] has suggested that

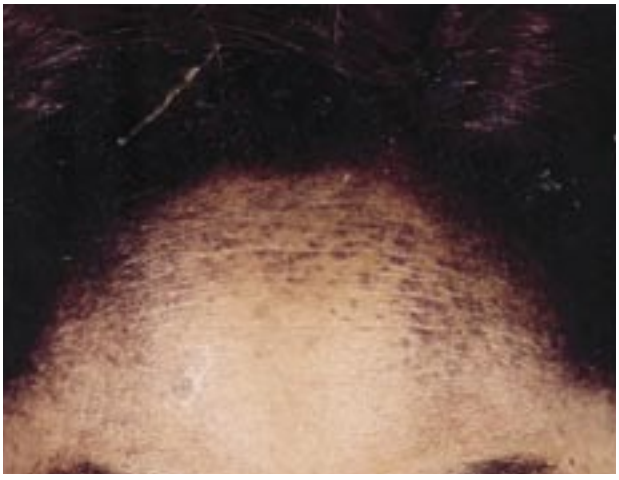


Figure 1. Diffuse large plaques with a reticulate peripheral pattern involving the upper back, upper forehead, and behind the ears.

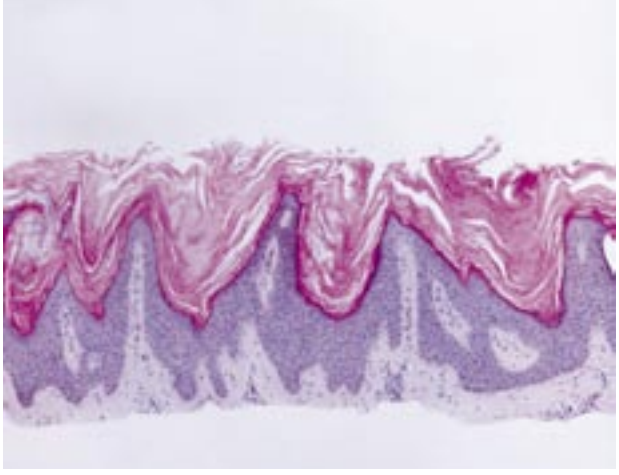


Figure 2. Skin biopsy of the papules on the back showed papillomatosis and hyperkeratosis (H&E, original magnification ×100). Periodic acid-Schiff stain was negative for fungal elements.

bacteria may play a role in the pathogenesis of the disease, where the bacteria or its products (bacterial superantigens) induce the epidermal proliferative changes. This could explain the response to antibiotic treatment. However, to date, no bacteria have been isolated in CRP.[15] Other theories include photosensitivity[16]; genetic factors[3,4]; amyloidosis cutis[17]; and endocrine disorders, such as diabetes mellitus and thyroid disease, but all of these correlations are weak. Gougerot and Carteaud[18] suspected an endocrine cause; however, most reported patients have been healthy. Groh et al[17] hypothesized CRP to be an early form of amyloidosis cutis, but they have been the only authors to find amyloid in the lesions.

Many therapies are available for CRP, but none have been universally effective. Certain antibiotics (eg, tetracyclines and macrolides) have been effective in many patients, though it is uncertain if this is attributable to anti-inflammatory immunomodulatory effects or a direct antibiotic effect.[14,19,20] Oral or topical retinoids (eg, systemic etretinate or isotretinoin, topical tretinoin 0.1%) also have been successful.[9-11] Recently, topical calcipotriol, a vitamin D analogue, has been used with a favorable response and few side effects.[6,12,21,22] The effectiveness of retinoids and vitamin D_3 analogues further supports the theory that CRP is caused by a defect in keratinization because both regulate cell differentiation.[23]

REFERENCES

1. Gougerot H, Carteaud A. Papillomatose pigmentee innominee. *Bull Soc Fr Dermatol Syphilol.* 1927;34:719.
2. Angeli-Besson C, Koeppel MC, Jacquet P, et al. Confluent and reticulated papillomatosis (Gougerot-Carteaud) treated with tetracyclines. *Int J Dermatol.* 1995;34:567-569.
3. Baden WP. Familial cutaneous papillomatosis. *Arch Dermatol.* 1965;92:394-395.
4. Henning JP, de Wit RF. Familial occurrence of confluent and reticulated papillomatosis. *Arch Dermatol.* 1981;117:809-810.
5. Faergemann J, Fredriksson T, Nathorst-Windahl G. One case of confluent and reticulate papillomatosis (Gougerot-Carteaud). *Acta Derm Venereol.* 1980;60:269-271.
6. Carrozzo AM, Gatti S, Ferranti G, et al. Calcipotriol treatment of confluent and reticulate papillomatosis (Gougerot-Carteaud syndrome). *J Eur Acad Dermatol Venereol.* 2000;14:131-133.
7. Champion RH, Burton JL, Burns DA, et al, eds. *Textbook of Dermatology.* Osney Mead, Oxford: Blackwell Science; 1998.
8. Roberts SO, Lachapelle JM. Confluent and reticulate papillomatosis (Gougerot-Carteaud) and *Pityrosporum orbiculare*. *Br J Dermatol.* 1969;81:841-845.
9. Baalbaki A, Malak JA, al-Khars MA. Confluent and reticulated papillomatosis. Treatment with etretinate. *Arch Dermatol.* 1993;129:961-963.
10. Bruynzeel-Koomen CA, de Wit RF. Confluent and reticulated papillomatosis successfully treated with aromatic etretinate. *Arch Dermatol.* 1984;120:1236-1237.
11. Hodge JA, Ray MC. Confluent and reticulated papillomatosis: response to isotretinoin [letter]. *J Am Acad Dermatol.* 1991;24:654.
12. Ginarte M, Fabeiro JM, Toribio J. Confluent and reticulated papillomatosis (Gougerot-Carteaud) successfully treated with tacalcitol. *J Dermatolog Treat.* 2002;13:27-30.
13. Lee SH, Choi EH, Lee WS, et al. Confluent and reticulated papillomatosis: a clinical, histopathological, and electron microscopic study. *J Dermatol.* 1991;18:725-730.
14. Jang HS, Oh CK, Cha JH, et al. Six cases of confluent and reticulated papillomatosis alleviated by various antibiotics. *J Am Acad Dermatol.* 2001;44:652-655.
15. Montemarano AD, Hengge M, Sau P, et al. Confluent and reticulated papillomatosis: response to minocycline. *J Am Acad Dermatol.* 1996;34:253-256.
16. Vassileva S, Pramatarov K, Popova L. Ultraviolet light-induced confluent and reticulated papillomatosis. *J Am Acad Dermatol.* 1989;21:413-414.
17. Groh V, Schnyder UW, Sigg C. Papillomatose papuleuse confluente et reticulee. Gougerot-Carteaud—a further form of skin amyloidosis? *Dermatologica.* 1981;162:118-123.
18. Gougerot H, Carteaud A. Neue Formen der Papillomatose. *Arch Dermatol Syphil.* 1932;165:232-267.
19. Humbert P, Treffel P, Chapuis JF, et al. The tetracyclines in dermatology. *J Am Acad Dermatol.* 1991;25:691-697.
20. Fung MA, Frieden IJ, LeBoit PE, et al. Confluent and reticulate papillomatosis: successful treatment with minocycline. *Arch Dermatol.* 1996;132:1400-1401.
21. Gulec AT, Seckin D. Confluent and reticulated papillomatosis: treatment with topical calcipotriol. *Br J Dermatol.* 1999;141:1150-1151.
22. Kurkcuoglu N, Celebi CR. Confluent and reticulated papillomatosis: response to topical calcipotriol. *Dermatology.* 1995;191:341-342.
23. Griffiths CE. Gougerot-Carteaud still an enigma after all these years. *J Dermatolog Treat.* 2002;13:27-30.