Coexisting confluent and reticulated papillomatosis and terra firma-forme dermatosis

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Abstract
Confluent and reticulated papillomatosis (CARP) is an uncommon skin disorder that usually affects young adults. The clinical presentation most often involves the upper trunk, commonly in the intermammary area or, less frequently, the interscapular and epigastric regions.1,2 The lesions usually appear as grayish brown, hyperkeratotic, verrucous papules and patches that are confluent centrally and reticulated peripherally. While usually asymptomatic, the eruption can present with mild pruritis.1 In addition, the eruption is often fairly symmetrical across the body and trunk. We present an unusual case of CARP coexisting with terra firma-forme dermatosis in a young male, with atypical features consisting of a significant unilateral distribution with a whorled appearance.

Introduction
Confluent and reticulated papillomatosis of Gougerot and Carteaud (CARP) was first described in 1927. This condition typically has an onset during puberty, and is usually sporadic although familial cases have been reported. Young women are affected 2.5 times more frequently than young men, and blacks are affected twice as often as whites.1

There are several different theories surrounding the pathogenesis of this skin disorder. Some theorize underlying endocrine imbalance and insulin resistance, due to an association with diabetes, obesity, pituitary and thyroid disorders.1,3 However, others theorize a keratinization defect or an association with Malassezia furfur, due to successful treatment with retinoids and selenium sulfide, respectively.1

Terra firma-forme dermatosis is characterized by dirt-like plaques, occurring most often in children or adolescents, that cannot be washed away with soap and water but can be removed with isopropyl alcohol. Differential diagnosis often includes CARP, pityriasis versicolor, and acanthosis nigricans.4,5 Misdiagnosis of this condition often leads to unnecessary workups including skin biopsies and endocrine evaluations.

Case Report
A 15-year-old male of Middle Eastern descent initially presented to our dermatology clinic with his mother with a four-year history of an unresolving rash. He noticed gradual progression of the lesions across his neck, chest and back. He denied any associated symptoms or precipitating factors, including recent infection, fever, chills, joint aches, pain or pruritis. Past medical history and skin history were noncontributory. Cutaneous examination revealed hyperpigmented, brownish-black, warty papules and plaques, predominantly unilateral, distributed across his right anterior neck, right chest, bilateral arms, epigastric skin, and back in a whorled appearance. The anterior chest revealed a vertical linear demarcation, with the right chest affected to a greater extent than the left (Figures 1, 2). At this time, the patient refused a biopsy as well as lab work. We initiated treatment with tacrolimus ointment 0.1%, which he found to provide mild improvement of dyspigmentation.

The patient then presented a year later stating that the lesions were not improving to his satisfaction and he would like to pursue more aggressive treatment. We ordered a comprehensive metabolic panel, lipid panel, antinuclear antibody, complete blood count, erythrocyte sedimentation rate, urinalysis, hemoglobin A1C and carcinoembryonic antigen, which were all within normal limits. A biopsy of the epigastric skin was also done at this time, which revealed an epidermis with slight acanthosis with papillomatosis and hyperkeratosis, and pityrosporum yeast in the cornified layer (Figure 3). We initiated further treatment with doxycycline 100 mg

Figures 1 and 2. Hyperpigmented, brownish-black, warty papules and plaques, predominately unilateral and with a whorled appearance.

Figure 3. Histopathology showing the epidermis with slight acanthosis with papillomatosis and hyperkeratosis, and pityrosporum yeast in the cornified layer.
orally once daily. At two-week follow-up, the doxycycline and topical tacrolimus had resulted in minimal improvement, and we therefore discussed other treatment options such as isotretinoin, oral minocycline, hydroquinone and antifungals. The patient refused further treatment and decided to continue with the doxycycline and topical tacrolimus. At this visit it was also discovered that some of the hyperpigmented lesions could be wiped off with gauze soaked in isopropyl alcohol, indicating a coexisting diagnosis of terra firma-forme dermatosis (Figure 4).

Discussion
CARP was first described in 1927. It is most common in people in their late teens and early 20s. CARP affects all racial groups but has been found to occur more frequently in blacks. The lesions begin as round, red-to-brown papules, 1 mm to 2 mm in diameter, distributed across the intermammary region and, less commonly, the interscapular and epigastric regions. As the disorder progresses, the lesions enlarge to 4 mm to 5mm and can become hyperkeratotic and verrucous. The lesions can coalesce, and the eruption tends to spread centrifugally, becoming confluent centrally and reticulated peripherally.1,2

In our case, the lesions appeared as hyperpigmented, brownish-black papules and plaques distributed across the intermammary, epigastric and interscapular regions. The presentation of this eruption was unique due to its whorled appearance and unilateral distribution across the anterior chest. Due to these unique clinical features, our differential diagnosis was broad and included acanthosis nigricans, progressive cribriform and zosteriform hyperpigmentation, and linear and whorled nevoid hypermelanosis. Both clinically and histologically, this case resembled both acanthosis nigricans and CARP, but CARP was favored due to the distribution throughout the trunk as opposed to limited to intertriginous areas. Progressive cribiform and zosteriform hyperpigmentation was ruled out based on the presence of lesions on the bilateral back and bilateral arms. We also ruled out linear and whorled nevoid hypermelanosis due to the lack of lesions present in infancy, as well as the lack of epidermal melanosis on histology.6,4 While the diagnosis of terra firma-forme dermatosis was made at a subsequent visit, we feel this was a secondary diagnosis to the CARP. Co-occurrence of these two conditions has previously been reported, and distinguishing them can be difficult as they can present clinically and histologically alike.5,8

The papillomatous skin changes associated with CARP may trap debris, dirt, or sebum, resulting in terra firma-forme dermatosis.9 Treatment of CARP has proven difficult, as it often does not respond to therapeutic interventions or recurs after therapy has stopped. Many treatments have been reported to achieve success, such as a variety of antibiotics, isotretinoin, acitretin, salicylic acid, hydroquinone, antifungals and 5-fluorouracil, though no single therapy has been proven to be consistently successful.1,2 In our case, our patient had mild improvement of dyspigmentation with topical tacrolimus and oral doxycycline, but the treatment did not result in full, desired improvement. While lesions of terra firma-forme dermatosis can be treated successfully with cleansing with isopropyl alcohol, continued maintenance treatments may be necessary when an underlying papillomatous condition such as CARP is also present.

Conclusion
While CARP has many characteristic clinical features, there have been a variety of different manifestations reported.2 We report another manifestation of CARP, presenting primarily unilaterally with a whorled pattern and with the coexistence of terra firma-forme dermatosis. Familiarity with terra firma-forme can avoid unnecessary workups, but it is also important to recognize that papillomatous diseases such as CARP and acanthosis nigricans can possibly predate patients to this entity. When CARP or acanthosis nigricans is suspected, it may be prudent to first attempt wiping off hyperpigmented lesions with isopropyl alcohol at initial visit. However, as in the case of our patient, in the presence of terra firma-forme it is important to closely examine the skin for textural surface changes after lesions have been wiped away, because they may suggest a co-existing papillomatous disease.

References

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