

Dermoscopic Features and Diagnostic Criteria of Galli-Galli Disease: A Clinical Update

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Introduction

Galli-Galli disease (GGD) is a very rare acantholytic disease belonging to the reticulate pigmentary disorders, a group of cutaneous diseases mainly inherited as autosomal dominant [1]. Sporadic cases of GGD have been described [2]. The clinical resemblance with Dowling-Degos disease (DDD) and the presence of the same gene mutations in DDD [3] prompts considering GGD and DDD as two distinct phenotypes of the same pathological entity.

In GGD mutations in genes *KRT5*, *POFUT1*, *POGLUT1*, or *PSENEN*, affecting melanosome transfer and melanocyte and keratinocyte differentiation, may account for the polymorphous phenotype. While *KRT5* mutation is responsible for classic and generalized DDD and “classic” GGD, mutations in *POGLUT1* are described in the generalized “atypical” GGD phenotype, in which the characteristic

flexural lesions are absent and persistent macules are distributed mainly on trunk and extremities [4].

Case Presentation

Case 1. A healthy 41-year-old Caucasian female presented with a 2-year history of a slightly pruritic rash affecting the trunk. Physical examination revealed thin flat non-folliculotropic papules, some of which scaly, admixed with mottled lentigo-like macules (Figure 1A). The flexural areas and the face were spared. Dermoscopy of papular lesions showed a yellowish-to-brown-tan star-like or polygonal central area surrounded with a whitish halo on a faint erythematous background with fine linear-curved vessels at the periphery (Figure 1B). Macular lesions, on the other hand, showed a pseudo-reticular pattern with well demarcated borders (Figure 1C). At histological

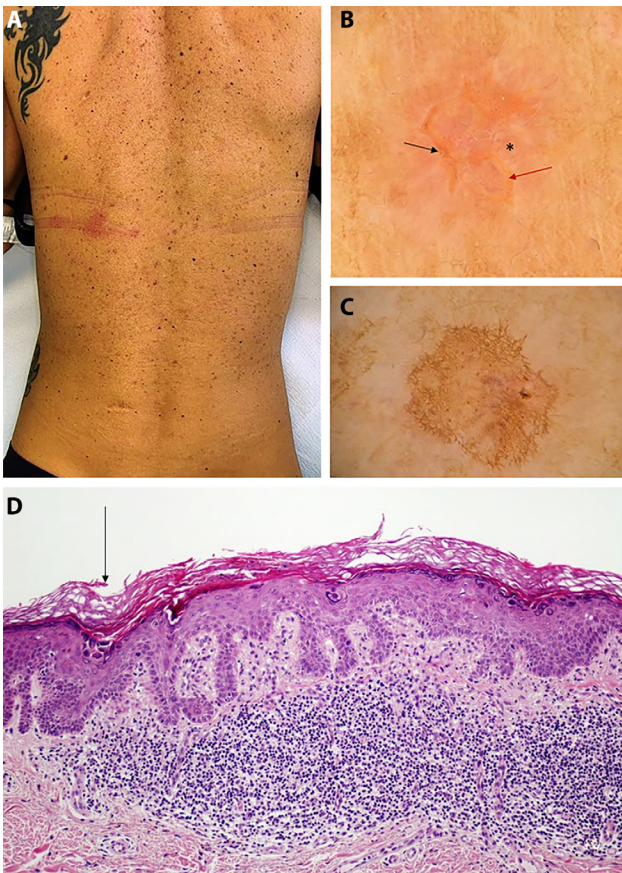


Figure 1. (A) Papular and macular lesions on the trunk in a 41-year-old female. (B) Dermoscopy of a Galli-Galli disease (GGD) papule with a polygonal yellowish center (black arrow), a whitish halo (red arrow), and fine linear and curved vessels at the periphery (asterisk). (C) Dermoscopy of a macule with pseudo-reticular pattern and clear-cut borders. (D) Histology of a GGD papular lesion showing subtle focal acantholysis in the stratum spinosum and granulosum (arrow) with rare dyskeratotic keratinocytes (H&E, original magnification $\times 100$).

examination of a papule, elongation of the rete ridges, hyperparakeratosis, and deceptively focal acantholysis in the stratum spinosum and granulosum with occasional dyskeratinocytes were present (Figure 1D). The dermis showed a moderate lymphocytic infiltrate. Based on the clinical findings and on the histopathological features, the patient was diagnosed with GGD.

Case 2. A 50-year-old female presented with a 10-year history of crops of asymptomatic fine erythematous-brownish papular lesions limited to the lower limbs associated with round lentigo-like macules (Figure 2A). Dermoscopic findings of the papular and the macular lesions were similar to those described in Case 1, but in Case 2, macular lesions prevailed, and “hybrid” papulo-macular lesions were

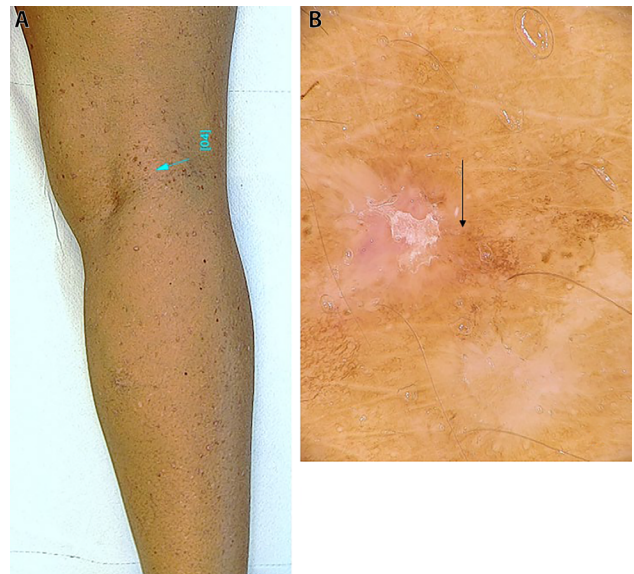


Figure 2. (A) Papular and macular lesions of the limb of a 50-year-old female. (B) Dermoscopy of an almost involute papular lesion showing a star-like uniform pinkish center with scales fading to macules with a pseudo-network pattern (black arrow).

observed (Figure 2B). Histology of a papular lesion showed the same characteristic as observed in Case 1.

Conclusion

In GGD, an accurate histopathological examination is necessary to identify foci of acantholysis as this clue may be extremely subtle in the atypical, generalized variant, and it is mainly linked to the age of the lesion. Of note, as acantholysis is commonly observed in different dermatoses (Table 1), clinical-pathological correlation is always mandatory.

To our knowledge, dermoscopic features of GGD have been only reported by two groups [5,6], which also employed reflectance confocal microscopy and line-field optical coherence tomography, with a good correlation to histological features, suggesting a reliable non-invasive approach. Thus, the whitish central area and the lentigo-like pattern observed with dermoscopy likely correspond respectively to the hyper-parakeratosis and acanthosis and to the elongate rete ridges seen at histology.

The dermoscopic brownish star-like brown pattern with a thin peripheral halo and a pinkish background, along with the absence of follicular plugging and inclusion microcysts, are very characteristic of GGD. Moreover, we showed a possible dermoscopic continuum between the papular and the macular late phase.

Table 1. Papular Acantholytic/Dyskeratotic Disorders Clinically Mimicking GGD.

Disease	Diagnostic clues
Dowling-Degos disease (DDD)	Genetically inherited. Hyperpigmented macules in a reticulate distribution. Dermoscopy may show follicular plugging and milia within papular lesions.
Grover disease	Itchy papular and vesicular eruption on trunk and proximal extremities. Superficial indentations/micro fissures at dermoscopy.
Darier disease	Genetically inherited with variable expressivity. Warty itchy lesions on seborrheic areas of the trunk and face. Palmoplantar and nail manifestations.
BRAF-inhibitor-induced acantholytic dyskeratosis	Oncological patient under treatment.
Acantholytic dyskeratotic acanthoma (acantholytic dyskeratoma)	Solitary lesions. Superficial indentations/microfissures at dermoscopy.

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