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Case Report

An acantholytic variant of dowling degos disease

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ABSTRACT

Galli- Galli disease is a rare acantholytic variant of Dowling- Degos disease, with few cases reported in the literature till date. We here, describe an interesting case of Galli- Galli disease with prominent histopathological findings. Future studies and genotype-phenotype correlations may elucidate why acantholysis is observed in some patients, but not in others.

1. Introduction

Galli- Galli disease is a rare acantholytic variant of Dowling-Degos disease (DDD), with histological finding of focal acantholysis. It was first reported by Bardach et al. in 1982 [1] who described the disease in two brothers. [2] It has an autosomal dominant pattern of inheritance.

Dowling-Degos disease is a skin condition characterized by a lacy or net-like (reticulate) pattern of abnormally dark skin pigmentation, particularly in the body's folds and creases. These skin changes typically first appear in the armpits and groin area and can later spread to other skin folds such as the crook of the elbow, back of the knee, and under the breasts. Less commonly, pigmentation changes can also occur on the neck, wrists, back of the hands, face, scalp, scrotum, and vulva. These areas of hyperpigmentation typically cause no health problems. [3]

Individuals with Dowling- Degos disease may also have hyperpigmented lesions on face, comedon- like lesions on back and neck, pitted perioral scars and rarely palmar pits. Fluid-filled sacs within the hair follicle may develop, most commonly on the scalp. Rarely, affected individuals have patches of skin that are unusually light in color (hypopigmented). [4]

In rare cases, individuals with Dowling-Degos disease experience itching (pruritus) or burning sensations on the skin. These feelings can be triggered by UV light, sweating, or friction on the skin. [1,5]

The pigmentation changes characteristic of Dowling-Degos disease typically begin in late childhood or in adolescence, although in some individuals, features of the condition do not appear until adulthood. [6] New areas of hyperpigmentation tend to develop over time, and the other skin lesions tend to increase in number as well. The skin changes associated with Dowling-Degos disease may cause distress and anxiety in individuals.

2. Case Report

A 29 year old female, known case of hypothyroidism on treatment presented with widespread, asymptomatic to mildly itchy lesions on body present since 1 and half year. There is history of similar complaints in her younger sister. The skin lesions began on thighs and then gradually spread to involve trunk, bilateral flanks and bilateral forearms. Clinical examination revealed numerous generalized, symmetrically distributed brownish macules and few pruritic erythematous papules present over bilateral

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thighs and trunk. The face, palms and soles were spared and she didn't have any nail, hair, teeth, flexure and mucosal involvement. Dermoscopy from the lesion showed scaling, brownish pigmentation in the background along with crater like areas. On histopathology, increased basal layer pigmentation along with downward elongation of rete ridges was seen. Focal area of prominent suprabasal non-dyskeratotic acantholysis was also present.[3] Dermis showed few melanophages with mild perivascular lymphocytic infiltrate.Figures 1, 2 and 3

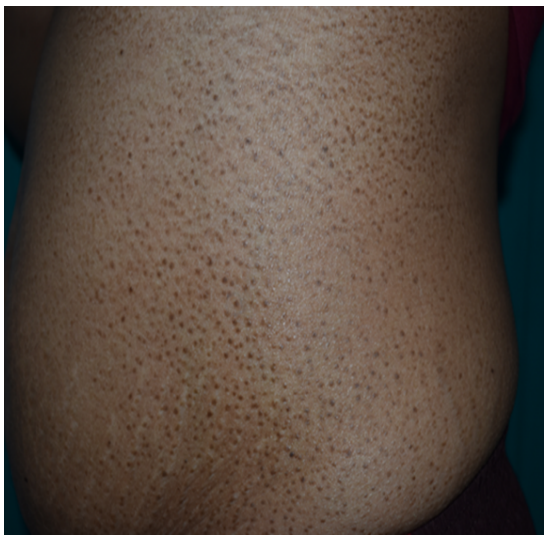

Fig. 1: Multiple well defined hyperpigmented to brownish papules and macules present over abdomen, bilateral flanks

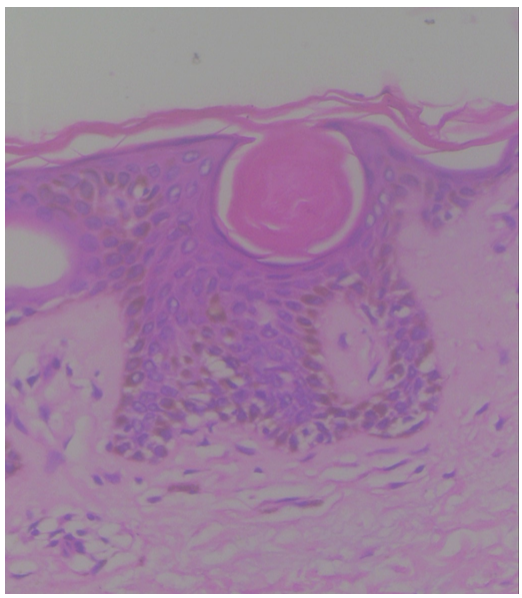

Fig. 2: Histopathology showing- digitiform elongation of the rete ridges along with suprabasal focal acantholysis

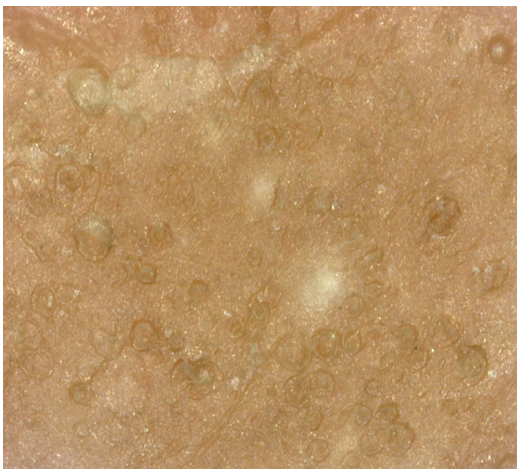

Fig. 3: Dermoscopy from lesion showing several crater like areas on brownish background, along with mild scaling

3. Discussion

Galli-Galli disease is a benign genodermatosis. Its mode of inheritance is believed to be autosomal dominant with variable penetrance, but it can occur sporadically. As with DDD it is believed to be linked to mutations in the KRT5 gene along with mutation in POFUT1, POGLUT1 and PSENEN genes[7] which affects notch signaling pathway. The disease usually presents with hyperpigmented macules in the skin folds. Other findings include papules, scaly erythematous plaques, comedo-like lesions and pitted perioral scars. In our case, hyperpigmented macules and a few papules were present; however, there was no involvement of flexures which makes this case more interesting and unique. Histopathologic examination reveals digitiform elongation of the rete ridges seen in DDD, together with suprabasal focal acantholysis.[8] There are around 10 reported[9] cases of Galli-Galli disease in literature in which males were more commonly involved than females, age group commonly involved was between 40-50years, flexural involvement was present in all of them with histopathology showing acantholysis along with dyskeratosis in few of them.[10] There are no case reports in literature mentioning about dermoscopic features of Galli-Galli disease, along with specific histological findings. Clinicopathological and dermoscopy, correlation plays a very important role in such cases for reaching upto a specific diagnosis and management of patient.

4. Conclusion

Our case report underlines clinicopathological correlation and dermoscopy plays a very important role in diagnosing such cases and providing better patient outcome.. Future studies and genotype-phenotype correlations may elucidate why acantholysis is observed in some patients, but not in

others.

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6. Conflict of Interest

None declared.

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