Generalized Asymptomatic Cutaneous Pits and Comedones in a Young Woman

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A 22-year-old female, born out of a non-consanguineous marriage, presented with multiple, asymptomatic and progressive dark papular and pitted skin lesions in a generalized distribution from the past 5 years. She was also informed about similar complaints from her father. Examination revealed discrete comedonal and shallow pitted lesions involving the face, trunk [Figure 1a-c] and extremities. Rest of the cutaneous and systemic exam was unremarkable. Polarized dermoscopy of the comedonal lesions revealed central blue-black structureless area surrounded by a white halo. The pitted lesions showed crateriform depressions with a surrounding white halo [Figure 1d]. Biopsy of a comedonal lesion showed dilated hair follicle containing lamellated keratin together with elongated and branched rete ridges of the infundibular and immediate perifollicular epidermis with increased melanization of the basal layer [Figure 2].

What is your diagnosis?

Answer

Follicular Dowling-Degos disease (DDD)

Discussion

The classical form of DDD has an autosomal dominant inheritance pattern that usually manifests around puberty and is characterized by symmetrically distributed reticular hyperpigmentation principally of the flexures. This pigmentary anomaly may be accompanied by comedonal and perioral pitted scars.[1] Variants of this disease include a generalized form with flexural and non-flexural distribution, a histological variant (Galli-Galli disease) and the rare follicular variant. The disease can also be associated with multiple epidermoid cysts, hidradenitis suppurativa and rosacea-like facial erythema (Haber syndrome).[2,3] Follicular DDD manifests with signs and symptoms confined to the hair follicles as described in the index case without any features of the classical form of the disease. In addition to mutations in keratin 5 gene, follicular abnormality has also been proposed in the pathogenesis accounting for the follicular lesions and association with hidradenitis suppurativa. Follicular DDD in association with ichthyosis vulgaris and hidradenitis suppurativa has been described as well. Chloracne (predominantly facial lesions), familial dyskeratotic comedones (larger lesions sparing

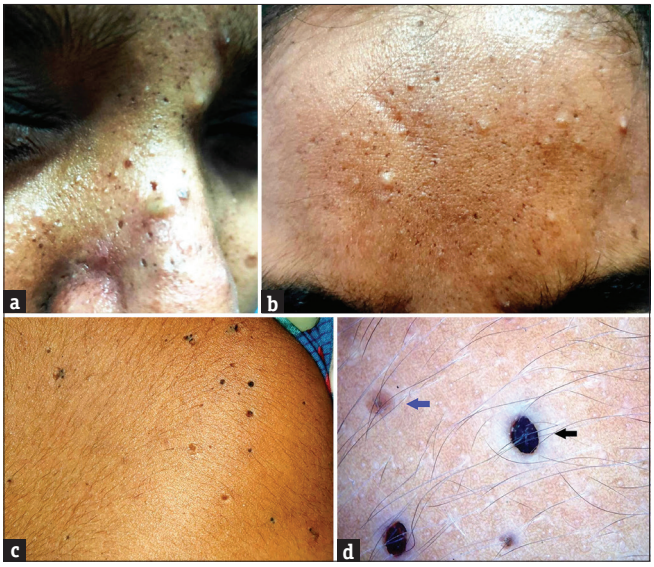

Figure 1: Multiple discrete comedonal and pitted lesions on the centrofacial area (a), forehead (b) and back (c). Polarized dermoscopy at x10 magnification (using DermLite™ DL3, 3Gen Inc., San Juan Capistrano, CA, USA) showing central blue-black structureless area with a white halo (black arrow) and crateriform depression with a white halo (blue arrow) (d)

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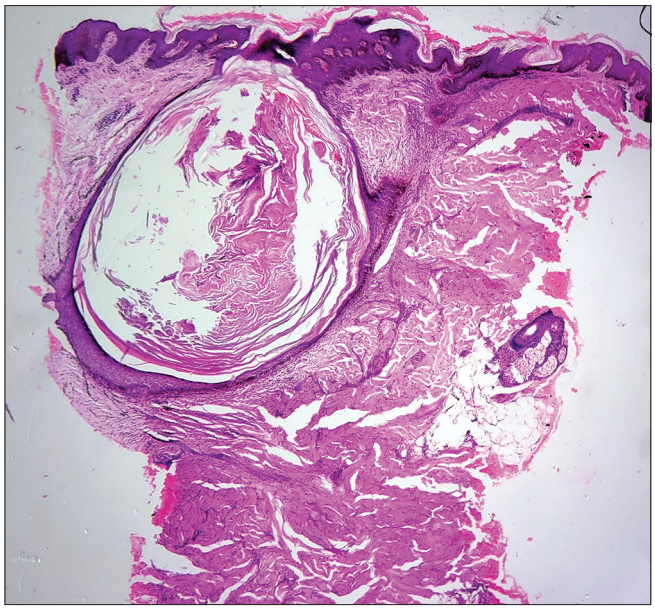

Figure 2: Photomicrograph showing dilated hair follicle containing lamellated keratin together with elongated and branched rete ridges of the infundibular and immediate perifollicular epidermis with increased melanization of the basal layer. [H and E, x10]

the head and neck region) and comedonal Darier disease (associated with nail, oral mucosal and other specific cutaneous manifestations) can mimic follicular DDD.[2-4] Treatment of follicular DDD is difficult. Topical and systemic retinoids for follicular lesions and laser resurfacing for pitted scars have been employed with minimal benefit.[3,5]

A diagnosis of follicular DDD was considered in the index case based on the history, clinical, dermoscopic and histological findings, and the case is presented to highlight the importance of recognizing this disorder as well as to differentiate from the other conditions with a similar presentation for appropriate management. Our patient was made aware of the nature of the disease and absence of any specific and curative treatment for the same. For cosmetic betterment, the patient was offered fractional carbon dioxide laser resurfacing for the facial pitted scars. The patient, however, was lost to follow-up.

Learning Points

- Follicular DDD is a rare variant of the classical form of the disease with the signs and symptoms confined to hair follicles.
- Histology of follicular DDD shows features similar to that of the classical disease but limited to the follicular infundibular epithelium.
- An awareness of this rare follicular variant of the disease is useful for appropriate diagnosis and management of the disease as well as in differentiating from other conditions with similar clinical presentation

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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