Follicular Dowling–Degos Disease: A Rare Pigmentary Dermatosis

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Dowling–Degos disease (DDD) is a rare autosomal-dominant genodermatosis characterized by reticulate pigmentation of flexures. We are reporting two cases of DDD of follicular variety, a mother and her daughter, from a family in which 10 out of 16 individuals (five males and five females) were affected.

A 32-year-old female with her 12-year-old daughter presented with discrete, tiny, hyperpigmented, keratotic, follicular papules all over the body, hyperpigmented macules along with comedo-like lesions over the face, and atrophic pitted scars over the buttocks, thighs, and periumbilical area [Figures 1-3]. The daughter had lesions over bilateral cubital fossae and popliteal fossae and very few lesions over the face, neck, and legs without any scarring [Figure 4]. Lesions started developing at around puberty in both the cases. Family history revealed the involvement of 10 out of 16 individuals [Figure 5]. Dermatoscopy of a comedo-like lesion showed thick, hyperkeratotic, hyperpigmented follicular plug. A skin biopsy from a hyperkeratotic, hyperpigmented papule revealed large keratinous plug in the infundibulum and focal areas of thin, branching, pigmented epithelia with a downward proliferation of infundibular wall. The interfollicular epithelium was not involved [Figure 6]. A diagnosis of follicular DDD was made on the basis of prominent follicular nature of the lesions, which was supported by family history, physical examination, histopathological, and dermatoscopic findings. Therefore, in cases of pigmented genodermatoses, such as DDD, histopathological examination is important to differentiate the disease entity from other pigmented dermatoses and to classify it as a variant of the disease.

DDD presents with numerous hyperpigmented macules in a reticulate pattern over flexural areas. Its association with comedo-like papules, perioral pitted scars, epidermoid/trichilemmal cysts, chloracne-like lesions, hidradenitis suppurativa, seborrheic keratoses, palmar pigmentation, and rosacea-like lesions (Haber's syndrome) have been reported in the literature.[1,2] It has many variants [Table 1], out of which the follicular variant is very rare; till now very few cases of pure follicular DDD have been reported in the literature.[3,4]

Singh *et al*. first reported two cases from two different families (from India) of follicular DDD with characteristic histopathology showing follicular restriction of the lesions, as seen in our cases.[4] Few cases with follicular hyperpigmented macules and pitted scars in combination with reticulate

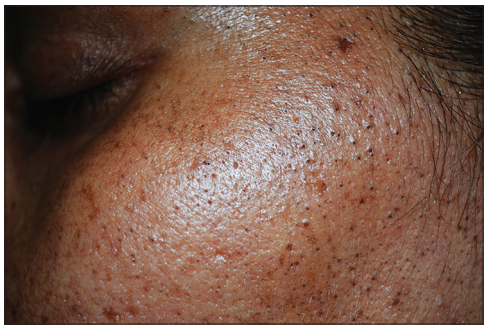

Figure 1: A photograph of the left cheek showing multiple hyperpigmented, hyperkeratotic papules, hyperpigmented macules and comedo-like lesions

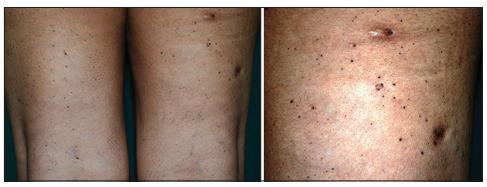

Figure 2: A photograph of the thighs showing hyperpigmented keratotic papules with pitted scars

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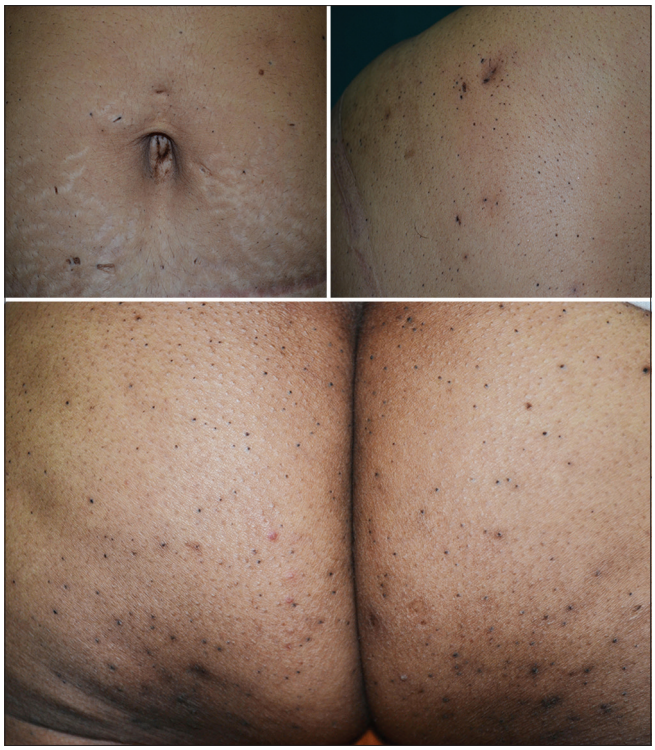

Figure 3: A photograph showing comedonal papules and pitted scars over the abdomen, buttocks, and upper back

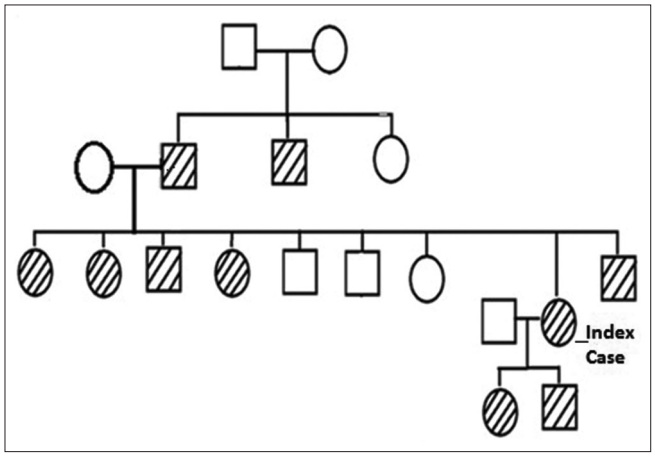

Figure 5: The family tree showing autosomal dominant inheritance

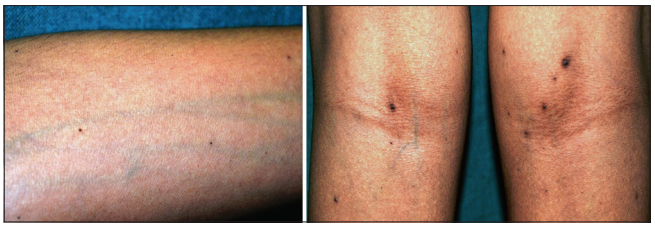

Figure 4: Photograph of the daughter showing keratotic papules over the forearm and popliteal fossae

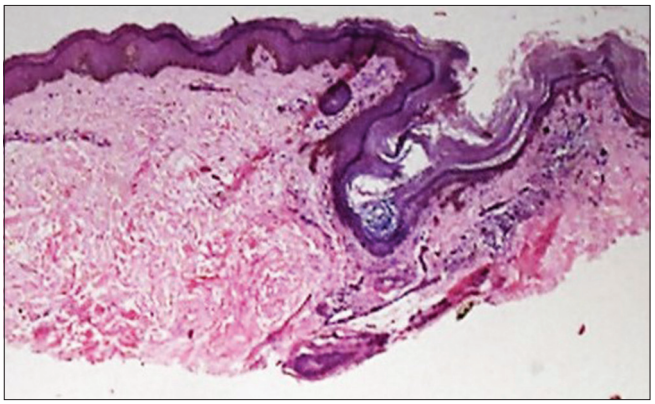

Figure 6: Skin biopsy from a comdonal papule revealed large keratinous plug in infundibulum and focal areas of thin, branching, pigmented epithelia with the downward proliferation of infundibular wall (H and E, ×40)

Table 1: Variants of DDD

Variant of DDD	Characteristics
Classic DDD	Reticulate hyperpigmentation in flexural areas
Generalized DDD	Hyperpigmented or hypopigmented papules and macules in generalized distribution
Galli-Galli disease	Histopathological variant with acantholytic cells
Follicular DDD	Hyperpigmented, hyperkeratotic papules restricted to follicles
Haber's syndrome	Pigmented keratotic papules on the axilla, neck, and torso with pitted scars on the face and persistent facial erythema
Pigmentatio Reticularis Faciei et Colli	Hyperpigmentation of the face and neck with multiple epidermoid cysts

hyperpigmentation have been reported, however, the isolated follicular variant of DDD has been rarely reported.[1]

Genetically there is a loss of functional mutations in the keratin 5 gene.[5] However, recently, several reports have highlighted the possible role of follicular pathology in its genesis. Indicators of this follicular pathology include clustering and punctate nature of the lesions, association with hidradenitis suppurativa, and comedo-like lesions, as well as prominent infundibular changes on histology.[4] Still there is a need for detailed research and more studies to understand the exact pathomechanism of this follicular variety of DDD.

This presentation intends to share information about this very rare genodermatosis and helps to differentiate the follicular variant of DDD clinically and histologically.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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