CASE 1

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Received: November 30, 2007
Accepted: January 10, 2008

Case

A 40–year-old woman referred to the dermatological clinic of Razi hospital with painful lesions on the axillae, between her breasts and intergluteal region since adolescence. The patient had been treated with a variety of antibiotics with only minimal improvement. The lesions reappeared with cessation of treatments. On cutaneous examination, multiple furuncle-like discharging nodules were observed over the axillae, between the breasts, buttock and gluteal region (figure 1). Other findings were giant black comedones and depressed scars which were found between the aforementioned lesions, particularly on her back (figure 2). A number of pits with 2 to 3 mm diameter were seen around the mouth (figure 3). Additionally, there were hyperpigmented macules in a reticular pattern over the axillae, inframammary area and groin (figure 4). Mucosal surfaces and nails were not pigmented. Systemic examination was normal. Laboratory data were all within normal limits. In familial history, one of her sisters, aged 50 years, had a similar pattern of pigmentation from the age of 20.

What is your diagnosis?

Move on to next page for the answer and discussion.

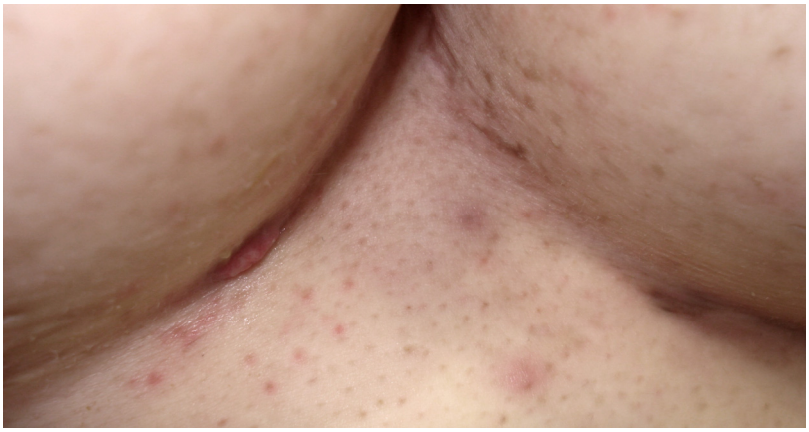

Figure 1

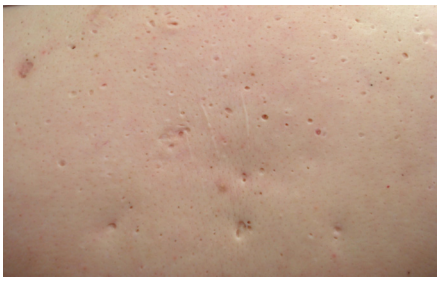

Figure 2

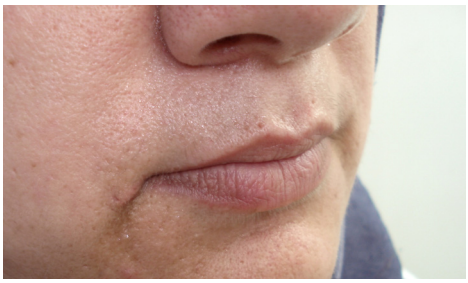

Figure 3

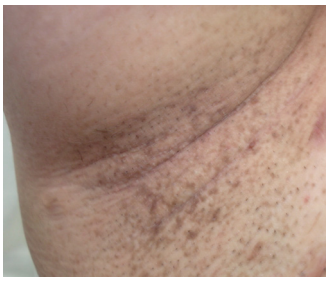

Figure 4

Diagnosis: Dowling-Degos Disease

Discussion

Dowling-Degos disease (DDD or reticulate pigmented anomaly of the flexures) is a rare genodermatosis, more common in women, with an autosomal dominant mode of inheritance.[1] It is mainly characterized by acquired reticulate pigmented macules of the flexural areas, inner aspects of the thighs and neck, and, rarely, wrists, face, scalp, scrotum, and vulva. The pigmentation is symmetrical, progressive and asymptomatic .Its color varies from brown to black. Other common features include pitted perioral acneiform scars and comedo-like dark hyperkeratotic follicular papules.[2-4]

The association of DDD and hidradenitis suppurativa is well documented. It is hypothesized that they could be due to clinical manifestation of a single underlying defect in follicular keratinisation.[5-7] Other less common manifestations of the disease are: epidermoid and tricholemmal cyst, multiple keratoacanthomas and pilonidal sinus.[5-8] Squamous cell carcinoma over the pigmented areas has been reported[9]. Some of these associations may have a pathogenic relationship with DDD8.

The etiology of the disease is unknown. It seems that a gene defect within the pilosebaceous unit and impaired desmosomal structures may play a role in its pathophysiology.[5,8]

The histopathological findings of the pigmented lesions include an acanthotic epidermis with irregular elongated rete ridges with basilar hyperpimentation on the tips. Involvement of the follicular infundibulum is noted. Also, thinning of the suprapapillary epithelium, moderate hyperkeratosis, dermal melanosis, perivascular lymphohistiocytic dermal infiltration and dermal fibrosis along with elongated rete ridges are observed.[2]

Some authors now believe that Dowling-Degos syndrome should be considered as a part of a diseases spectrum that includes reticulate acropigmentation of Kitamura, Haber syndrome and acropigmentation of Dohi. They may be variants of the same entity, with similar clinicopathological features and overlap cases.[4,8,10]

Treatment of Dowling-Degos disease is usually difficult and often disappointing. Transient good results are achieved with the use of topical corticosteroids, depigmentating agents, topical adapalene and Erbium-YAG laser.[11,12] Further studies including prolonged clinical evaluation and recognition of basic immunopathogenesis are required to evaluate the efficacy and safety of the different treatment options.

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