Hidradenitis suppurativa, Dowling–Degos and multiple epidermal cysts: a new follicular occlusion triad

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Summary

This case illustrates the rare association between hidradenitis suppurativa (HS) and Dowling–Degos disease (DDD). Furthermore the association of HS, DDD and multiple epidermal cysts has not to our knowledge been described before, but their coexistence in the same patient is likely to reflect the same follicular anomaly. It is possible that a single underlying defect of follicular proliferation may account for the coexistence of these conditions.

Report

A 49-year-old woman presented with recurrent nodules and abscesses in her axillae and groins since the age of 11 years. Interestingly, eight relatives from her father's family have similar although milder skin lesions. In addition, she noticed the appearance of unusual macular pigmentation in a flexural distribution and around her waistline, and three painful subcutaneous lumps, two on her thighs and another in her left groin.

Examination revealed inflammatory nodules and multiple sinuses in the groins (Fig. 1a). She also had comedones, dermal contractures and scars in the axillae and posterior neck. She had clusters of brownish macules in a reticulate pattern in her axillae, submammary areas, lower abdomen and groins (Fig. 1b).

Histology of excised skin from the posterior neck revealed follicular plugging and focal abscess formation, consistent with HS (Fig. 2a and b). Punch biopsy of a pigmented axillary macule showed elongation of the rete ridges, with concentration of melanin at the tips, supporting a diagnosis of DDD (Fig. 2c). Histology of one of her subcutaneous swellings on her thigh confirmed that they were epidermal cysts.

Our patient had a body mass index of 37 kg/m^2. Although obese, she was afraid to lose weight as previous attempts, albeit successful, had resulted in psychological disturbances. Nevertheless, she had lost 30 kg over the past 3 years but had not noticed any improvement in her HS. She had been taking Micronor, a progestogen-only pill, which may well have exacerbated her HS. She had not been a candidate for Dianette because she was a smoker. Smoking may be another aggravating factor in itself.

Multiple treatment modalities proved unsuccessful, including topical clindamycin, and prolonged courses of various antibiotics, singly or in combination. Isotretinoin and acitretin produced no benefits and had to be discontinued due to side effects.

A localized area of HS was excised twice. More recently she was commenced on spironolactone and cimetidine without any improvement. Despite a trial of finasteride, injection of isolated lesions with intralesional corticosteroids and replacement of Micronor with a more oestrogenic contraceptive preparation, her HS remained recalcitrant to treatment.

HS is a chronic, suppurative and cicatricial disorder of follicular differentiation affecting apocrine-rich cutaneous sites. DDD which is characterized by reticulate pigmentation, follicular plugging and pitted scars, is also a disorder of follicular derivation. HS may be associated with DDD.[1] Although not generally regarded to be of genetic origin, family studies of patients with HS support the possibility of it being a single gene disorder with an autosomal dominant pattern of inheritance.[2] Indeed, there is a strong family history of this disorder illustrated in our case. Our patient and her family have been

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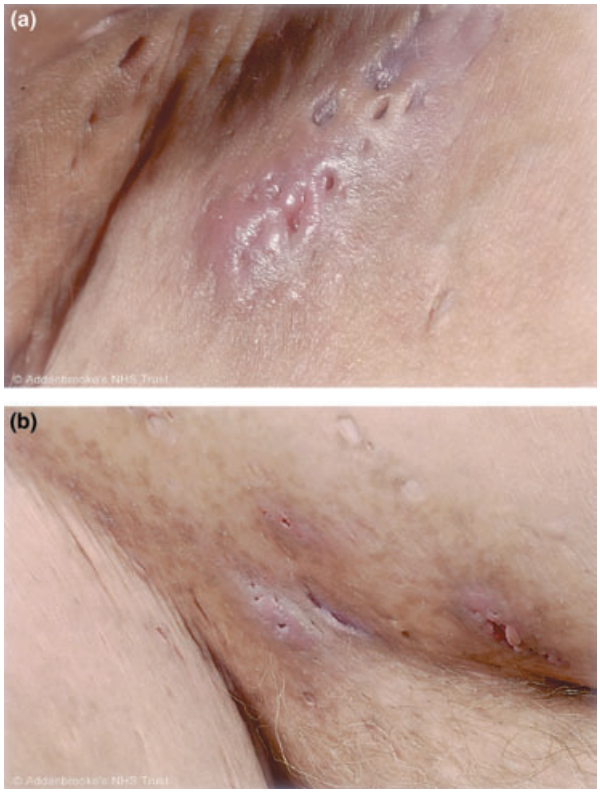
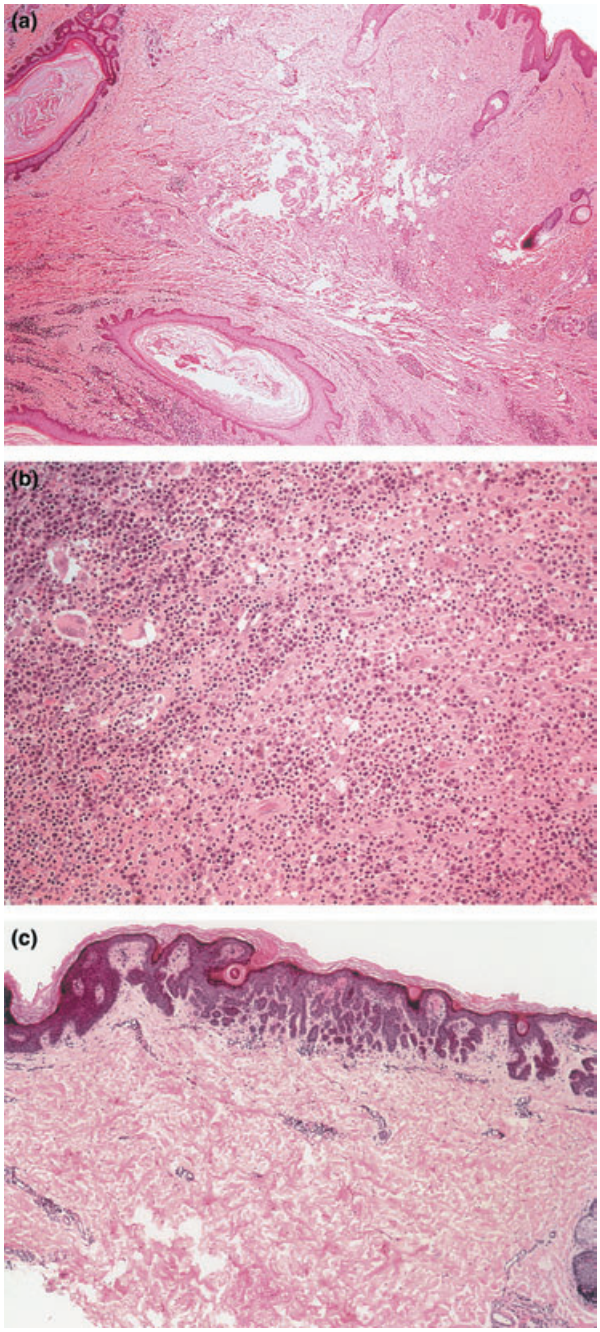


Figure 1 (a) Inflammatory nodules, sinuses and abscesses in the groin area. (b) Reticulate and macular hyperpigmentation of DDD.

referred for investigation with regards to any underlying genetic abnormalities for their condition. DDD is also an autosomal dominant condition.[3] It is proposed that genetic abnormalities of the hair follicles are shared by the two conditions.[4] However, due to the lack of phenotypically similar appearances between the two disorders, it seems more likely that the follicular anomaly inherent in DDD may predispose to the development of HS.

The follicular occlusion triad of HS, acne conglobata and dissecting cellulitis of the scalp is of course well recognized. However, the triad of HS, DDD and multiple epidermal cysts, as seen in our patient, has not been reported in the literature. Nevertheless, it is not surprising for these three conditions to occur together since all share the same follicular anomaly.

HS is a chronic relapsing and remitting condition and current therapies consist mainly of empirical attempts to control the disease. There are only three randomized controlled trials. These included a double-blind cross-over trial of cyproterone acetate,[5] topical clindamycin vs. placebo[6] and topical clindamycin vs. systemic tetracycline.[7] Systemic antibiotics, hormonal therapy and corticosteroids may be effective but the common



Figures 2 (a, b) Histology showing follicular plugging, chronic inflammation and focal abscess formation, consistent with HS (haematoxylin and eosin; original magnification ×40).
(c) Acanthosis and elongated rete ridges of the follicular epithelium together with increased melanin pigment at the tips, diagnostic of DDD (haematoxylin and eosin; original magnification ×40).

experience is one of initial response followed by subsequent relapse when such treatment is withdrawn. Intralesional corticosteroids are a good option for

isolated inflamed lesions. Retinoids appear to be of limited value, acitretin being more useful than isotretinoin. There are also anecdotal reports of the successful treatment of HS with ciclosporin, dapsone, infliximab and finasteride.[8]

Surgery is essential in many cases, especially for localized, recurrent, refractory and advanced disease. Incision and drainage carries a recurrence rate of 100% while wide excision and surgical reconstruction has a recurrence rate of 25% at a median interval of 20 months. Careful laying open of sinus tracts and healing by secondary intention may be a good compromise. Carbon dioxide laser therapy provides an alternative efficient method for surgical removal of chronic fistulating lesions of hidradenitis.[9]

Many different treatment options have been tried for DDD without convincing therapeutic benefit. Topical adapalene was reported to be effective in one case report.[10] The Er:YAG laser treatment led to good clinical results in another case.[11]

In summary, a 49-year-old woman with recalcitrant HS, DDD and multiple epidermal cysts is discussed. The constellation of HS, DDD and multiple epidermal cysts in a single patient has not, to our knowledge, been reported previously. It seems likely that a single underlying defect in follicular epithelial proliferation may account for the coexistence of these clinically distinct disorders of follicular occlusion.

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