Hidradenitis suppurativa. An update

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SUMMARY

Hidradenitis suppurativa is a chronic and often disabling disease characterized by intermittent periods of inflammation and abscess formation in apocrine sweat gland-containing skin. Various therapies have been implemented in its treatment with variable results. Pathogenesis, histopathology and therapy of this disease are reviewed.

KEY WORDS

hidradenitis suppurativa, follicular occlusion, review

Introduction

Hidradenitis suppurativa (HS) is a chronic disease of unknown etiology that mainly affects the genitofemoral, perianal and axillary regions (1-4). The disease is believed to be follicular in origin (1). HS has a higher prevalence in women than men and is estimated to affect 4 percent of women in the general population (5).

Etiology

HS often occurs in multiple members within families, suggesting a genetic predisposition (5). Various etiological agents have been associated. The increased incidence of HS in obese women with acne led to the theory of HS being associated with an hyperandrogenic endocrine disorder (6,7). One study did not find evidence for biochemical hyperandrogenism in women affected with HS (8). Thus, the role of androgens in HS is still not clear. Smoking (9), lithium (10) and oral contraceptives (11) may also be associated with HS, possibly as triggering factors. Nonetheless, the etiology of the disease is unknown.

Pathogenesis

Historically, apocrinitis was believed to be the defining histologic feature and pathogenetic mechanism of HS (12,13). Characteristic lobular abscesses in the apocrine gland, demonstrated histologically, seemed to further implicate apocrine gland involvement in the manifestation of disease (12-14). Recently, histological evidence points to HS being a follicular disease (1,15-19). Histological examination in the majority of speci-

mens reveals follicular involvement, including poral occlusion and folliculitis (1). Apocrinitis as the dominant histological feature is found in only a small number of specimens (1,17). Furthermore, a paucity of apocrine glands was demonstrated in the genitofemoral region, one area commonly affected by HS (1). This finding supports the theory that apocrine gland inflammation is not the etiological and pathogenetic mechanism of HS, but rather a secondary manifestation of follicular involvement (1).

Clinical features

HS may arise singularly or multifocally in the genitofemoral, perianal and axillary areas (1-4). The lesions are painful and have a foul odor attributed to bacterial colonization. Erythematous dermal abscesses form that measure up to 2 cm in diameter. Untreated abscesses will gradually increase in size and may drain to the surface. The course of HS is chronic and remitting, with new abscesses arising in previously unaffected areas or in regions of past involvement. Scarring, fibrosis and sinus tract formations are manifestations of late disease. Strictures can occur secondary to sinus tracts; fistulas may also complicate HS. Squamous cell carcinoma is a rare sequella of longstanding HS (20-24). These cancers may be locally aggressive with distant metastases and a high mortality rate (25).

Clinical associations

The follicular occlusion triad consists of HS, acne conglobata and perifolliculitis capitis abscedens et suffodiens (26-28). Arthritis of peripheral joints and the axial skeletion may rarely be associated with HS (29-32). HS has also been linked to Crohn's disease (33-35). One study reported 24 out of 61 patients with HS were also diagnosed with Crohn's disease, which predated the HS by an average of 3.5 years (35). Acanthosis nigricans and Fox-Fordyce disease may predispose to HS (36). Pyoderma gangrenosum (37,38) and pyoderma vegetans (27) have also been associated with hidradenitis suppurativa.

Therapy

Treatment of hidradenitis suppurativa is challenging. Late stage disease, evidenced by the formation of sinus tracts, fibrosis and scarring, usually necessitates surgical intervention. Early HS is often best treated with antibiotics in our experience, although few clinical trials are available (39). Three months' of treatment with topical clindamycin decreased the number of abscesses, inflammatory nodules and pustules in twenty-seven patients with chronic HS (40). Systemic tetracycline therapy has shown similar clinical effectiveness (39). Others and also we often recommend intrealesional corticosteroids in early stage disease (41). The use of cyproterone acetate and ethinyl estradiol achieved successful clinical results in four women with chronic HS (42); clinical improvement with cyproterone acetate and ethinyl estradiol was also described in another study (43). Isotretinoin is only slightly effective in controlling the disease; clinical improvement is seen in patients with mild HS (44). The clearing of chronic, refractory perianal HS was seen after treatment with cyclosporin for concomitant pyoderma gangrenosum (38).

Medical therapy is of limited value once HS has progressed past its early stage (2). The surgical option of choice for late stage HS is wide local excision with healing by secondary intention (41). One study examined patients with chronic HS who had undergone surgery between the years of 1976 and 1997 (4). An estimated 72-month follow-up revealed that 45% of the patients had recurrence of local HP (4). A 100% recurrence rate was reported after drainage procedures, while limited and wide local excision techniques had a recurrence rate of 42.8% and 27%, respectively (4). More recently, carbon dioxide laser excision has been proposed as a better alternative to conventional surgery (2,45). Carbon dioxide laser excision offers better hemostasis and visualization of abscessed tissue than conventional surgical techniques, allowing more accurate excision (2).

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