

Recognizing and Managing Comorbidities and Complications in Hidradenitis Suppurativa

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■ Abstract

The list of comorbidities associated with hidradenitis suppurativa (HS) is extensive, although these diseases do not necessarily share a common causality. Among the categories of comorbidities that are observed are obesity, other skin diseases, inflammatory conditions, and genetic disorders. Complications include scarring, restricted movement resulting from scarring and fibrosis in underlying tissue, conditions associated with obstructed lymph drainage, and psychosocial issues. Adverse effects on quality of life are common and may be severe, including unemployment, deterioration of family and other social relationships, and suicidal ideation. Clinical intervention for HS must include consideration and attention to these comorbidities and complications.

Semin Cutan Med Surg 33(supp3):S54-S56

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■ Keywords

Comorbidities; dermatologic diseases; hidradenitis suppurativa; immune-mediated inflammatory diseases; obesity

Hidradenitis suppurativa (HS) is a disease that must be considered and treated in a context much broader than just the consideration of skin lesions and sinus tracts. Clinicians who diagnose HS also must recognize and be prepared to manage the significant comorbidities and complications associated with this chronic, debilitating disease.

Comorbidities and HS

The comorbidities associated with HS have been widely discussed in the literature and include both common (**Table**)¹ and rarely occurring associations. However, the term “association” is used here with the caveat that, in most instances, the exact nature of

the association between HS and its comorbidities is unclear. The diseases are comorbid with respect to observed coexistence, but not necessarily with respect to a common causality. As future studies reveal additional information about the genetics of all chronic diseases, genetic associations and links will become more clearly defined. Meanwhile, it is not yet known whether one or more genetic defects exist that predispose individuals to both HS and the other frequently associated comorbidities. In addition, it is important to note that associations between HS and some conditions observed to be comorbid are not strongly supported by statistics from studies of large patient populations.

Obesity

The comorbidity most commonly associated with HS is obesity, with or without the other features that characterize the metabolic syndrome (hypertension, hypertriglyceridemia, low high-density lipoprotein levels, increased fasting blood sugar concentrations). Reported percentages of patients with HS who were overweight or obese range from slightly more than 50%^{2,3} to 75% or more in some older studies.⁴

Obesity certainly can exacerbate the symptoms of HS through several mechanisms, including increasing the area of skin-to-skin contact and promoting increased sweating and occlusion. In addition, hormonal changes associated with obesity—resulting in androgen excess—has been proposed as a possible inciting factor, involving changes in the hair shaft that promote follicular occlusion.⁵

However, beyond these associations, it is not known whether obesity is linked genetically to a predisposition for HS or just serves as a trigger for expression of the disease in susceptible individuals.

Comorbid Skin Conditions

HS is associated with numerous skin diseases, especially those categorized as follicular occlusion conditions (acne conglobata, dissecting cellulitis of the scalp, and pilonidal cyst^{6,7}) and acne vulgaris.¹ In addition, other, less commonly seen comorbidities include acral pustular psoriasis, acanthosis nigricans, and pyoderma gangrenosum.¹ Recently, a report was published suggesting that HS may result in systemic amyloidosis.⁸

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Publication of this CME article was jointly provided by the University of Louisville School of Medicine Continuing Medical Education and Global Academy for Medical Education, LLC, and is supported by an educational grant from AbbVie, Inc.

Dr Menter has received an honorarium from Global Academy for Medical Education for his participation in this activity. He acknowledges the editorial assistance of Joanne Still, medical writer, and Global Academy for Medical Education in the development of this continuing medical education journal article. Joanne Still has no relevant financial relationships with any commercial interests.

Alan Menter, MD, has been a consultant and/or investigator and/or speaker and/or advisory board member for AbbVie, Allergan Inc., Amgen Inc., ApoPharma Inc., Boehringer Ingelheim, Celgene Corporation, Convoy Therapeutics Inc., Eli Lilly and Company, Genentech, Janssen Biotech, Inc., LEO Pharma, Merck & Co., Inc., Novartis Pharmaceuticals Corporation, Pfizer Inc, Symbio/Maruko, Syntrix Biosystems, Wyeth, and XenoPort, Inc.

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■ TABLE Diseases Commonly Associated With Hidradenitis Suppurativa¹

Diseases of follicular occlusion (follicular occlusion triad)

- Acne vulgaris
- Acne conglobata
- Dissecting cellulitis of the scalp

Pilonidal cyst

Crohn's disease

Obesity/metabolic syndrome

Inflammatory Conditions

Patients with HS frequently experience symptoms of arthritis and arthralgia, noted particularly during HS flares and in association with acne conglobata.¹ Immunologic associations between these conditions have been suggested but, to date, have not been clearly established.

For more than 2 decades, case reports and studies involving relatively small numbers of patients have appeared regarding a possible association between Crohn's disease and HS. In some, Crohn's disease occurred prior to the onset of HS; in others, HS symptoms appeared first. Church and colleagues⁹ reported that, in all such cases, HS lesions developed in the perineal and perianal areas, but patients frequently also had concomitant HS lesions in the axillae, groin, and buttocks. More generally, inflammatory bowel diseases, as a group, are commonly associated with HS.^{10,11}

HS also has been reported as a comorbid condition in patients with several rare syndromes, including SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) and PAPA (pyogenic arthritis, pyoderma gangrenosum, and acne) syndromes.

Genetic Disorders and HS

Clinicians with practices containing a large number of patients with HS have on occasion seen patients with genetic disorders including Down syndrome and KID (keratitis, ichthyosis, and deafness) syndrome. The existence of these genetic comorbidities raises the issue of possible common sources of disease expression, but also raises questions about why certain individuals with conditions such as the metabolic syndrome—a clear comorbidity for HS—develop HS, whereas others do not, and why certain patients with HS tend to have disease that progresses slowly and seems to remain at the Hurley stage II level, whereas others progress to stage III, sometimes in a rapid course.

Complications of HS

The most common complications associated with HS can be grouped into four categories: scars, restricted movement resulting from surface scarring and fibrotic changes in underlying tissue, obstructed lymph drainage, and psychosocial issues. In addition, case reports have been published describing the development of squamous cell carcinoma (SCC)¹² and other malignancies in patients with long-standing HS.

Scarring in HS is frequently cosmetically disfiguring, contributing to the psychosocial problems so common with this disease. Strictures of the anus, rectum, and urethra may occur secondary to HS lesions in the groin and genitourinary areas. Contractures from scarring and fibrosis in the axillae and groin, a complication of a long-standing disease process, can result in limited mobility of the arms and legs, respectively.

Severe lymphedema is a late complication resulting from scarring of the lymph glands and obstruction of lymph drainage.¹³ Cases of disfiguring and disabling genital swelling also have been reported, including scrotal elephantiasis.¹⁴⁻¹⁶

As described earlier, patients with HS of long duration have an increased risk for malignancy, particularly SCC.^{7,12} In a retrospective study involving more than 2000 patients, Lapins and colleagues¹⁷ found that half of the patients with HS had an increased risk for malignancy. With respect to SCC specifically, the risk for this type of cancer was increased by 4.6-fold among patients with HS¹⁷; men with HS are more likely than women to develop SCC.¹⁸

Psychosocial Issues

Embarrassment, social isolation, and depression are inevitable consequences of HS. The exudates—often foul-smelling—that drain from active lesions are difficult to conceal, as are the multiple sinuses, contractures, and scars of healed lesions (Figures 1–3).



FIGURE 1 Axillary Hidradenitis Suppurativa. This 53-year-old female patient has draining sinuses and scarring from previously healed tracts.

Photo courtesy of Alan Menter, MD.



FIGURE 2 Multiple Lesions in Hidradenitis Suppurativa. This 32-year-old male patient has comedones, inflammatory nodules, multiple draining sinuses, and scarring in the axillary region.

Photo courtesy of Alan Menter, MD.



FIGURE 3 Inframammary Hidradenitis Suppurativa. These multiple draining nodules and sinuses formed in the inframammary folds in this 23-year-old obese male patient. Significant scarring also is evident.

Photo courtesy of Alan Menter, MD.

In a study of quality-of-life (QOL) impairment in 61 hospitalized patients, Wolkenstein et al¹⁹ used validated QOL instruments to compare HS with other chronic skin diseases, including psoriasis, chronic urticaria, and atopic dermatitis. The reported impairment in patients with HS was substantially more severe, although the authors note that the 61 subjects were hospitalized (ie, at the severe end of the HS spectrum).

In a 1996 study, Jemec and colleagues²⁰ reported that, overall, patients with HS lost an average of 2.7 workdays per year, but those with severe disease were unable to work at all because of permanent disabilities. In an early study attempting to characterize the specific psychosocial implications of HS, Anderson and coworkers²¹ reported that five of six patients in their study of patients with HS in the perineum, buttocks, and groin were unemployed and poor, and were divorced or reclusive. In an even earlier study, Anderson and Perry²² observed that axillary HS was associated with increased rates for unemployment, poverty, family deterioration, and suicidal ideation. More recently, Matusiak and colleagues²³ described the adverse effects of HS on QOL and professional activity, and Kurek et al²⁴ studied sexual health in patients with HS and reported that the adverse effects were “profound.”

Intervention for Comorbidities and Complications

With the exception of obesity, it is not known whether the treatment of comorbid conditions affects the development or course of HS; conversely, effective management of HS may have no implications for comorbid diseases. Weight loss certainly can decrease the risk for disease progression and can be an essential adjunct to disease-specific treatment. The reduction of skin-fold area can decrease friction, sweating, and mechanical occlusion of follicles, all factors that contribute to the development of HS lesions. In addition, other comorbid conditions that are part of the metabolic syndrome (eg, diabetes) will also be ameliorated by appropriate diet, weight loss, and even gastric bypass surgery in the morbidly obese patient.

Early identification and effective treatment of HS can prevent or mitigate complications of scarring, fibrotic changes, obstruction of lymph drainage, and the sequelae associated with these complications.

In addition to prompt and effective medical management of the physical aspects of the disease, clinicians can and should provide education, support, and practical information that can help patients cope with the psychosocial consequences of HS. Such efforts are easier to incorporate in dermatology, gynecology, family medicine, and primary care clinician practices than in the emergency care settings. However, because many patients with HS are seen in emergency departments, specialists in emergency medicine also can provide help in this area, despite the limitations of patient exposure time inherent in the emergency room.

Clinical experience shows that acknowledging to patients that their disease can be challenging and that support is available among others who are coping with HS can be comforting. Providing information, such as a patient education handout, and website information are enormously helpful. Strategies for coping with the psychosocial consequences of HS, including practical suggestions for managing employment and social challenges,²⁵ have the potential to change the negative psychosocial milieu that many of these patients endure.

Conclusion

HS has long been considered an orphan disease; however, evidence accumulated over the past decade has shown that the actual number of patients affected may be far greater than was previously known. Rather, HS has been a hidden disease, with many patients being identified only when the severity of symptoms drives them to seek the help of a medical practitioner—often in the operating room or in the hospital emergency department. Fortunately, HS has been the focus of therapeutic, academic, and research attention recently, and the unmet needs of patients with HS—in both improved medical therapy and attention to psychosocial issues—now are being addressed. Patient-focused support groups exist in many areas, with access to this type of assistance continuing to grow. Meanwhile, it is imperative that clinicians counsel patients and their families appropriately and, whenever possible, provide practical information such as printed handouts and contact information for patient support resources.

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