Natural History, Presentation, and Diagnosis of Hidradenitis Suppurativa

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Abstract

The diagnosis of hidradenitis suppurativa (HS) is based on a characteristic history and physical exam. The anatomic sites of involvement include the axillae (most common), groin, and buttocks, and the perianal, perineal, and mammary regions. Initially, HS manifests with open comedones (usually with two or more "heads") and tender subcutaneous acneiform papules. Without intervention, the natural history of HS is chronic and progressive. More painful subcutaneous nodules form, which rupture and drain a thick, mucopurulent, foul-smelling fluid. Later, sinus tracts form, and, over time, ropelike fibrotic subcutaneous scarring occurs, which can lead to disabling contractures of the affected limbs. Clinically, the severity of disease is classified using the Hurley staging system, which provides guidance for choosing among treatment options.
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Keywords

Follicular occlusion; hidradenitis suppurativa; Hurley staging system; Sartorius staging system

The diagnosis of hidradenitis suppurativa (HS) is a clinical one based on characteristic history and physical exam. No confirmatory laboratory tests exist, and although histologic features can support the diagnosis or rule out alternate diagnoses, histologic findings alone are not diagnostic. Therefore, skin biopsies generally are not recommended.

Natural History and Clinical Presentation

HS most commonly is seen in patients in the second and third decades of life. The disease is rare in prepubertal children, and onset of the disease is less common later in life. The most common sites of involvement are the axillae, groin, buttocks, and perianal, perineal, mammary, and inframammary areas, with the axillae most commonly affected. Lesion distribution varies by sex: The most common sites of involvement in women are the inframammary, axillary, and inguinal areas; in men, perianal HS and involvement of atypical sites such as the nuchal scalp and retroauricular areas occur more frequently (**Figure 1**). Although HS is three times more common in women than in men,[1] men tend to have more severe disease.

HS begins with follicular occlusion, followed by inflammation and, ultimately, rupture of the pilosebaceous unit. HS manifests with tender, subcutaneous, inflammatory nodules that resemble furuncles; these lesions generally are the first to come to medical attention. Acneiform papules and open comedones with two or more "heads" (double comedones) are also typical (**Figure 2**). When they first appear, inflammatory papules or nodules of HS frequently tingle, burn, and are associated with increased sweating. In obese patients with HS, multiple open comedones or double comedones may appear in intertriginous regions, likely resulting from increased areas of skin-on-skin contact, occlusion, friction, and rubbing. Patients often report flaring associated with sweating (as in warm weather or after prolonged and intense exercise) or in areas where clothing is tight against the skin.

Without therapeutic intervention, the disease typically progresses to form more fluctuant and more painful, subcutaneous nodules that resemble large furuncles. Unlike typical furuncles, these lesions may lack characteristic central rupture and drainage, instead opening laterally and draining a thick, mucopurulent, foul-smelling fluid. Deep dermal abscesses may join and progress to form chronically draining sinus tracts colonized by mixed bacterial flora. Long-standing sinus tracts form palpable, ropelike fibrotic subcutaneous scars (**Figure 3**). Particularly severe or extensive sinus networks can extend into deeper tissue, including muscle, fascia, lymph nodes, and other structures, depending on anatomic location (for example, the urethra or rectum).

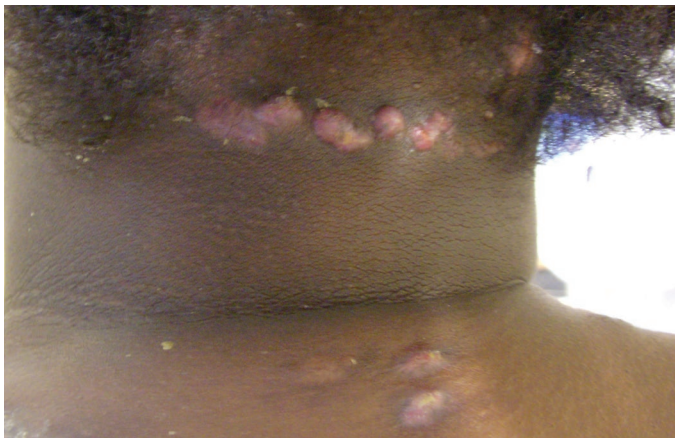

FIGURE 1 Hidradenitis Suppurativa Lesions of Posterior Scalp and Neck. This 23-year-old male patient presented with multiple inflammatory nodules in the scalp and neck area. This atypical anatomic presentation occurs more frequently in men than in women.
Photo courtesy of Alan Menter, MD.

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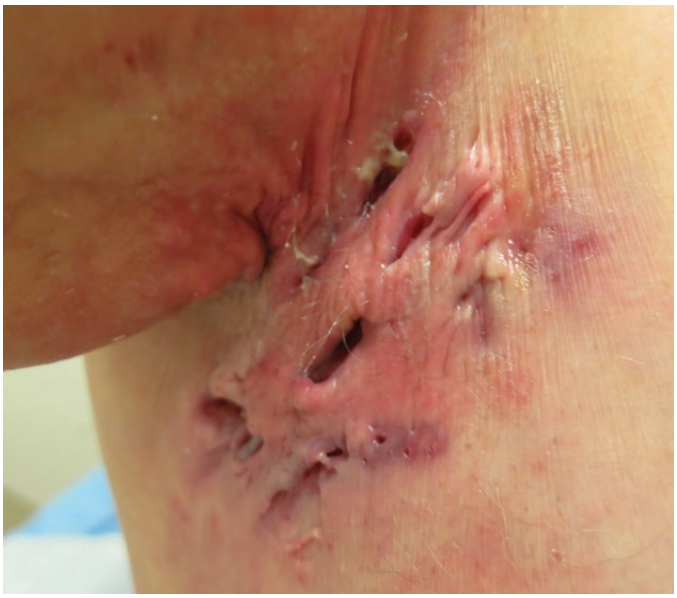

FIGURE 2 Severe Axillary Hidradenitis Suppurativa. The axillary region in this patient has active, draining sinus tracts that have formed ropelike, fibrotic subcutaneous scars.
Photo courtesy of Robert G. Micheletti, MD.

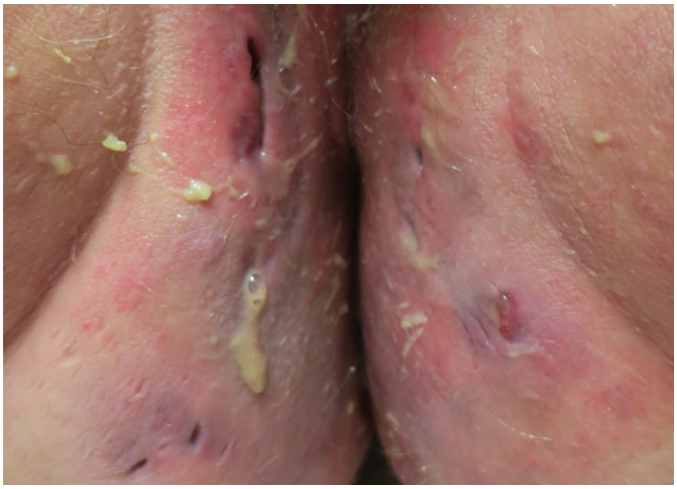

FIGURE 3 Severe Perineal Hidradenitis Suppurativa. Note the swelling, induration, and draining sinus tracts in the perineal region in this male patient.
Photo courtesy of Robert G. Micheletti, MD.

Establishing the Diagnosis

HS is a clinical diagnosis based on morphology and history, summarized by three key features. The diagnosis of HS can be made confidently if (1) the lesions are typical, (2) occur in the characteristic distribution, (3) and are chronic or recurrent. The onset of lesions usually is insidious, and patients typically are otherwise young and healthy.

The differential diagnosis for HS differs according to whether early or late lesions are present (**Table 1**). Among the list of possible diagnoses in patients with early HS lesions based on clinical appearance alone, the most common and most likely are furuncles, folliculitis, and atypically dilated comedones of acne vulgaris.

TABLE 1 Differential Diagnosis of Hidradenitis Suppurativa

Early lesions
• Acne
• Carbuncles
• Cellulitis
• Erysipelas
• Folliculitis
• Furuncles
• Inflamed epidermal inclusion cyst
• Lymphadenopathy
• Perirectal abscess
• Pilonidal cyst
Late lesions
• Actinomycosis
• Anal fistula
• Cat scratch disease
• Crohn's disease
• Granuloma inguinale
• Ischiorectal abscess
• Lymphogranuloma venereum
• Nocardia infection
• Noduloulcerative syphilis
• Pilonidal disease
• Tuberculous abscess

Distribution and anatomic location further narrow the clinical differential. Characteristic bilateral involvement of the axillae, perineum, or inguinal folds is suggestive of HS, whereas a single, unilateral nodule more likely represents a staphylococcal furuncle. Lastly, the appearance of characteristic lesions in characteristic locations in a recurrent or cyclic fashion, with onset, rupture, healing, and reappearance, suggests HS.

Unusual or atypical mimics of HS that could be considered include Bartholin's abscess in female patients as well as, in either sex, pilonidal cyst, granuloma inguinale and lymphogranuloma venereum, Crohn's disease, scrofuloderma, and other infectious or neoplastic causes of perforating lymph nodes.

Alikhan et al[2] proposed an algorithm for diagnostic workup based on four questions: (1) Is there more than a single inflamed lesion? (2) Is the course chronic with new and recurrent lesions? (3) Are the lesions bilateral? (4) Are the lesions located primarily in the milk line? A positive answer to all four questions indicates a diagnosis of HS. If the answer to any of these is negative, the authors suggest a systematic strategy for further investigation according to which feature is atypical.

TABLE 2 Hurley Staging System for Extent of Disease in Hidradenitis Suppurativa[3]

Stage I	Single or multiple abscesses, but no sinus tracts or scarring (cicatrization)
Stage II	Single or multiple separated, recurrent abscesses with tract formation and scarring
Stage III	Multiple interconnected tracts and abscesses involving an entire anatomic area

Histologic Manifestations

A biopsy is indicated to rule out an alternate disease process if HS is suspected but the clinical features are atypical. Histologic findings of HS depend on lesion age but may include follicular hyperkeratosis and plugging, follicular dilation and rupture, perifollicular lymphocytic or mixed inflammatory infiltrate, abscess formation, and sinus tracts with stratified squamous epithelium, foreign-body giant cells, and fibrosis.

Disease Staging

In 1989, Hurley[3] proposed a clinical staging system for characterizing the extent of disease in patients with HS (**Table 2**). This simple system has been used in clinical trials and as the basis for choosing a particular therapy. For example, for patients with stage I disease (abscess formation without sinus tracts and scarring), a treatment regimen that includes oral doxycycline, chlorhexidine wash, and application of a clindamycin solution may be sufficient to manage active lesions. In contrast, a patient with more severe stage II or stage III disease will likely require these measures plus more aggressive intervention to slow or arrest the progression of disease.

More recently, Sartorius and colleagues[4] proposed a comprehensive staging system that yields regional and total scores based on anatomic regions affected, the number and types of lesions, and the extent of involvement (ie, the distance between lesions and the presence of normal skin between lesions). Given its complexity, the Sartorius system is better suited to quantifying disease extent and improvement in a research setting than in clinical practice but may be superior to the Hurley staging system for the former purpose.

Importance of Early Diagnosis

The goals of diagnosis and treatment are to reduce the burden of disease, including pain, odor, and drainage, and to prevent disease progression by healing existing inflammatory lesions and preventing the formation of new lesions, permanent sinus tracts, scars, fibrosis, and contracture, with the goal of improving quality of life. Because Hurley stage I disease can be controlled well in most patients, whereas effective control of more severe stage II or stage III disease can be elusive, early diagnosis and treatment is essential. Canoui-Poitrine and colleagues[5] reported that 68.2% of patients with HS have Hurley stage I disease, 27.9% have stage II disease, and 3.9% have severe, stage III, disease. Unfortunately, patients with early HS may be less likely to seek medical attention when discomfort and other symptoms are tolerable and lesions are easily hidden by clothing. Most present when lesions rupture, discomfort worsens, or discharge becomes messy, foul-smelling, or embarrassing. Some patients fail to recognize the disease for what it is and avoid medical attention until it is advanced and symptoms are severe.

Providers may similarly fail to recognize and accurately diagnose HS. It is not uncommon for patients to be seen initially in emergency departments or urgent clinics for incision and drainage of "boils." Some patients return repeatedly to the emergency department for the temporary relief from discomfort that incision and drainage provide, until the diagnosis of HS is made and more useful and effective treatment is instituted for the disease.

Failure to diagnose HS accurately in stage I or II leads to unnecessarily prolonged patient discomfort and results in wasted expenditure of health care resources. Patient quality of life, including work and intimate relationships, may be severely affected during this time. HS is a highly distressing condition with an impact on quality of life as significant as any disease systematically studied in dermatology.[6] Effective treatment options exist that can slow or arrest the disease process when lesions are mild. Progressive disease, with fibrotic scarring, contractures, and loss of limb function, such as may be seen in the presence of severe axillary disease, is much more difficult to treat and may result in permanent disability.

Conclusion

In most cases, a clinical diagnosis of HS can be made on the basis of presentation: The lesions of HS are typical, occur bilaterally in a characteristic distribution, and are chronic and recurrent. Routine biopsies and other laboratory tests are not generally helpful in the diagnosis of HS, except when the presentation is atypical and they are necessary to rule out other diagnoses. Early diagnosis and institution of appropriate therapy can improve both physical and emotional well-being and quality of life. The sequelae of long-standing disease can be devastating and permanently disabling.

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