# Challenging Ulcerative Vulvar Conditions

## Hidradenitis Suppurativa, Crohn Disease, and Aphthous Ulcers

Kristen M.A. Stewart, MD

**KEYWORDS**

• Vulvar • Ulcer • Hidradenitis suppurativa • Crohn disease • Aphthous ulcers

**KEY POINTS**

- Hidradenitis suppurativa is a common condition that can be disabling and warrants early recognition and multimodal treatment.
- The vulvar lesions of metastatic Crohn disease present with a range of clinical findings and can precede the diagnosis of gastrointestinal symptoms by months to years.
- Aphthous ulcers are a clinical diagnosis of exclusion and can be classified as primary or secondary to systemic disease.

### HIDRADENITIS SUPPURATIVA

Hidradenitis suppurativa (HS), also known as acne inversa, and historically as Verneuil's disease, is a common, chronic, disabling disease that unfortunately is often misdiagnosed and undertreated.[1] HS is an inflammatory cutaneous disease localized to intertriginous areas, including axillary, inguinal, anogenital, and inframammary skin. It is characterized by acute, recurrent, painful cysts and draining nodules, as well as chronic secondary scarring, dyspigmentation, and fistula formation.[2] The disease presents after puberty, commonly during young adulthood, and has a duration of many years, with periodic improvement and worsening of symptoms. HS has an estimated prevalence of 1% to 4%,[3,4] with a notable female-to-male predominance of 3:1.[5] The clinical spectrum can vary in severity from relatively mild cases with occasional inflammatory papulonodules to severe cases with frequent inflammatory lesions, ulcerative draining sinuses, and severe scarring. Regardless of severity, this disease has a significant impact on a patient's quality of life, personal relationships, and self-esteem.[6–8]

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Total Dermatology Care Center P.A., 915 West Monroe Street, Suite 101, Jacksonville, FL 32204, USA
*E-mail address:* kstewart@totaldermcare.com

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The pathogenesis of HS has not been completely elucidated yet and is likely multifactorial, involving a complex interplay among multiple contributing factors: a primary dysfunction of the folliculopilosebaceous unit, the individual's genetics and hormones, and immune dysregulation. The current fundamental theory is that HS is a disease of the folliculopilosebaceous unit attributed to defects in the follicular wall. Influenced by an individual's genetics and hormones, keratinocytes in the follicular epithelium hyperproliferate and sebum is increased, causing occlusion of the follicle. The follicle expands, eventually ruptures, and releases sebum, skin cells, and bacteria, triggering an intense inflammatory response. In HS patients, this acute reaction does not heal normally, but ultimately transforms into a chronic wound. Current theory holds that, as the body attempts to heal the inflammatory response, it instead entraps the sebum, skin cells, and bacteria from the ruptured follicle as well as stem cells found in the pilar unit, creating a buried invasive proliferative gelatinous mass (IPGM) and thereby producing the cysts, sinus tracts, and draining nodules characteristic of the disease.[9–13]

Several factors seem to contribute to the pathogenesis of HS, including genetic predisposition, smoking, diet, obesity, hormones, bacteria, immune factors, and inflammation. The influence of genetic susceptibility in HS is significant, with about 40% of patients having an affected family member.[14,15] Smoking is considered to be a contributing factor to HS, because, unlike controls, the majority of HS patients are smokers or have a history of smoking,[3] and studies have shown that HS symptoms improve when smoking is discontinued.[16] This correlation is not surprising, because nicotine is a known trigger of inflammation and may contribute to follicular plugging.

The potential contributions of diet, obesity, and hormones seem to be interwoven. Theories are adapted from acne studies, which have proposed that high glycemic load diets and dairy consumption may trigger relative androgen excess by increasing the sensitivity of androgen receptors on the folliculopilosebaceous unit, thereby promoting sebum production and follicular plugging.[17] These same dietary triggers also contribute to obesity, which has been theorized to increase shear stress on intertriginous skin, further promoting follicular occlusion.[3,18,19] The apparent relationship between androgens and HS is demonstrated by disease onset at menarche, and by observations that symptoms flare for many women premenstrually or with exposure to androgenic progestins and often improve during pregnancy and after menopause.[15,20] Further, antiandrogen treatment shows some benefit for both men and women. Additional studies are necessary to delineate the roles of diet, obesity, and hormones in HS.

Bacteria have been implicated in the pathogenesis, and antibiotics have been used as a treatment—although not a cure—with some success. That HS is not cured with antibiotics supports the current understanding that the role of bacteria is secondary. Most early, nonfluctuant lesions tend to be sterile, whereas fluctuant and chronic lesions are often polymicrobial and culture a wide variety of bacteria, including staphylococci, streptococci, Gram-negative rods, and anaerobic bacteria.[21–23] Acutely, bacterial contaminants from normal skin likely contribute to HS by stimulating the inflammatory response. Scarring and entrapment of a bacterial biofilm may explain the chronic inflammation associated with HS and its lack of response to antibiotic treatment.[24] Positive cultures are seen more frequently in suppurative, fluctuant nodules and may represent overgrowth of normal flora secondary to infection.

Recent HS research has focused on the role of immune factors and systemic inflammation.[25,26] The cutaneous inflammation of HS only very rarely produces systemic symptoms, such as fever and lymphadenopathy. HS has, however, been associated with the metabolic syndrome, which is defined by hypertension, diabetes, dyslipidemia, and abdominal obesity.[25–27] The prevalence of the metabolic syndrome in