Hidradenitis Suppurativa

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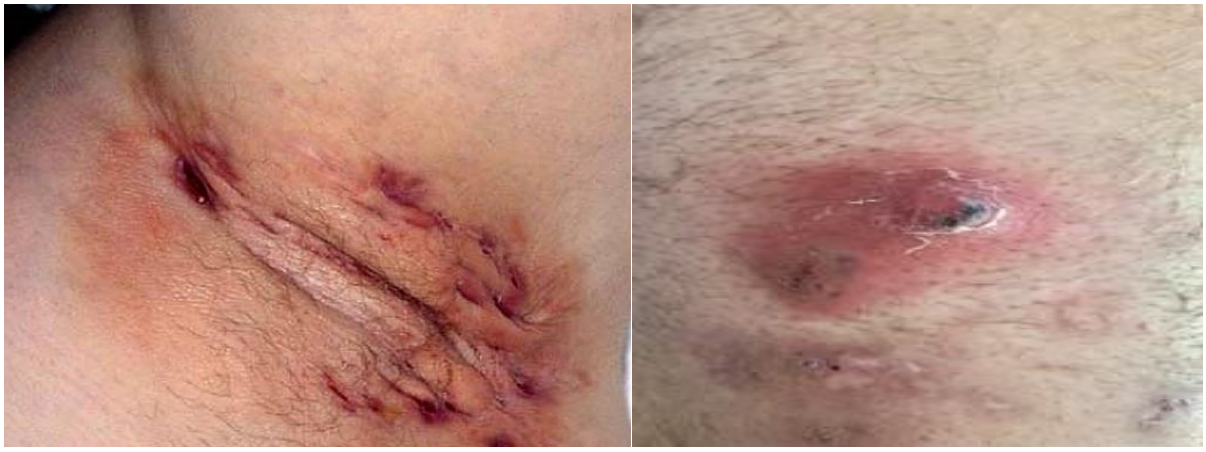
Hidradenitis suppurativa (HS) sometimes referred to as acne inversa, is a chronic, relapsing, inflammatory skin condition that typically occurs after puberty, with the average age of onset in the second or third decade of life. Patient with HS present with inflammation of hair follicles in the apocrine gland-bearing regions; armpits, genital area, groin, inframammary region, perianal region and buttocks that initially manifests as painful nodules or boils and progresses to abscesses, sinus tracts and scarring. [1, 2] Infrequently, hidradenitis suppurativa can occur in locations where apocrine glands are scant or absent and this is referred to as ectopic hidradenitis suppurativa.[6] The exact prevalence of HS remains unknown. Estimated prevalence is 1–4% worldwide, although these figures are thought to be underestimated because of under-reporting and misdiagnosis. HS is three times more common in women than men. [3]

The armpit and inguinofemoral regions are the most common locations for HS lesions in both sexes. Frontal lesions in the groin/thigh and breast tend to be more common in women, whereas lesions in the buttocks, perineal/perianal regions and atypical areas tend to be more common in men. [1, 2]



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Pathophysiology

The primary defect in hidradenitis suppurativa pathophysiology involves occlusion and subsequent inflammation of the hair follicle. A defective hair follicle becomes occluded and ruptures, emptying its contents, including keratin and bacteria, into the surrounding dermis. A chemotactic inflammatory response by surrounding neutrophils and lymphocytes can lead to abscess formation and subsequent destruction of the pilosebaceous unit and other adjacent structures. Other possible contributors to pathology include abnormal antimicrobial

peptides, abnormal secretion of apocrine glands, abnormal invaginations of the epidermis leading to tract formation, and deficient numbers of sebaceous glands.[2, 5]

Elevated levels of inflammatory cytokines including tumour necrosis factor alpha and various interleukins have been detected in the lesions of HS and provide possible targets for emerging treatments. Bacteria do not appear to be causative, as aspirate from non-ruptured lesions typically yields a sterile culture. However, bacterial infection and colonisation that occur during the process can secondarily worsen hidradenitis suppurativa.[2, 5]

Symptoms, Staging, Risk Factors and Comorbidities

The presentation of hidradenitis suppurativa is distinct, although the condition is not well-recognised except in dermatology clinics.[3] The most troublesome symptom of HS is chronic pain, of mild-to-moderate intensity, which is reported by almost all patients. The physical symptoms of hidradenitis suppurativa including, nodules or boils that progress to abscesses, sinus tracts and scarring, make it a painful condition. The pain associated with HS can be intense and chronic and is reported by patients as the most significant factor contributing to impaired quality of life. Up to 50% of patients report a burning or stinging sensation, pain, pruritus, warmth, and/or hyperhidrosis, 12–48 hours before an overt nodule occurs. Mean duration of a single painful nodule is 7–15 days. With time, the nodules may rupture, resulting in painful, deep dermal abscesses. With disease progression, draining sinus tracts, fibrosis, and scarring can be observed. After rupture, the lesions often extrude a purulent, foul-smelling discharge that stains clothing. HS is accompanied by embarrassment, social stigma, low self-worth and a negative impact on interpersonal relationships.[2, 5]

With reference to psychosocial evaluation, hidradenitis suppurativa can be a highly debilitating condition. The major factors influencing patients' well-being are disease severity, the number of flares or affected skin areas, and the lesion location. HS not only causes skin-related issues, but also has a profound impact on general quality of life measures, causing substantial deterioration of both physical and mental health. In both observational and registry studies, depression and anxiety were significantly related to HS. [4]

The Hurley Staging System is the simplest and most widely used instrument for HS classification in routine clinical practice. It classifies hidradenitis suppurativa into three stages: stage I: abscess formation, single or multiple, without sinus tracts and cicatrisation; stage II: recurrent abscesses with tract formation and cicatrisation, single or multiple, widely separated lesions; and stage III: diffuse or near-diffuse involvement, or multiple interconnected tracts and abscesses across the entire area. [5]

Severity is typically described according to the three Hurley categories. Most patients have grades I (mild) or II (moderate) hidradenitis suppurativa, with grade III (severe) disease reported in 4–22% of patients in recent studies. [1,

Hurley Staging System [1]

Grade I	Abscess formation, single or multiple, without sinus tracts and cicatrisation
Grade II	Recurrent abscesses with sinus tracts and cicatrisation; single or multiple widely separated lesions
Grade III	Diffuse or almost diffuse involvement, or multiple interconnected tracts and abscess across entire area

Risk factors for hidradenitis suppurativa include smoking, obesity, family history and other patient factors. Individuals with HS are commonly overweight or obese. Obesity leads to greater intertriginous surface area and skin friction, increased sweat production and retention, and hormonal changes leading to relative androgen excess, all of which are associated with HS. Metabolic syndrome is more common in obese individuals and is also seen more commonly in HS. Smoking is prevalent among those diagnosed with HS and it is thought that nicotine may cause increased follicular plugging. 33% - 40% of individuals with HS report an affected first-degree relative, suggesting a hereditary component with an autosomal dominant transmission pattern. In a small subset of affected families, researchers have identified a mutation of the gamma-secretase Notch signalling pathway.[2] The influence of hormones can also be seen in HS. There is a greater prevalence in females than males, with the age of primary occurrence most commonly between puberty and menopause. In addition, there are fluctuations of occurrence and severity with menstrual cycles and exogenous hormones. [3]

Comorbidities for hidradenitis suppurativa include inflammatory bowel disease, spondyloarthropathies, tumours and pyoderma gangrenosum. [1, 2] Patients with HS have a higher prevalence of gastrointestinal disease. The prevalence of IBD is 4–8 times higher in HS patients than in the general population, although there is no association with any distinct HS subtype. Compared to the general population, patients with IBD are 9 times more likely to develop HS.[5] Although the association is rare, it is speculated that pyoderma gangrenosum and HS share a common aetiology that may involve cytokine dysregulation. An increase of epithelial and non-melanoma skin cancers has also been reported in patients with HS. [3]

Diagnosis

Early diagnosis is very important for patients with hidradenitis suppurativa, in order to ensure the best possible course and prompt disease management. However, HS diagnosis generally occurs after an average 7-year delay, because the early stages are often mistaken for other conditions.[2, 5]

Diagnosis of hidradenitis suppurativa is made by history and clinical observation, and a biopsy is rarely needed. However, a biopsy is beneficial to rule out squamous cell carcinoma in the

presence of severe HS, if the diagnosis is uncertain. There are no blood tests to confirm HS and bacterial cultures are not beneficial unless secondary infection or an alternative diagnosis is suspected. Imaging is not typically required however, ultrasound may be a useful tool pre-operatively to identify the extent of sinus tracts. Lesions may warrant further imaging including MRI in severe perianal disease. HS can easily be differentiated from other conditions by the age of onset and the characteristic locations of lesions. Differential diagnosis includes, common abscess, carbuncles, furunculosis, infected bartholin's gland, Inflamed epidermal cysts, pilonidal cyst, scrofuloderma, actinomycosis, lymphogranuloma venereum and crohn's disease.[2, 5]

Treatment

Treatment choices for hidradenitis suppurativa are determined by disease severity and individual subjective impact. The degree of HS clinical involvement is usually determined according to the three-stage Hurley system.[2]

In early uncomplicated cases, topical antibiotics are the first line treatment. Topical clindamycin has been the most effective. Most significant effect is seen with superficial lesions while efficacy is poor with deep lesions. Intra-lesion corticosteroids can reduce local inflammation, and partial de-roofing/ punch debridement of individual lesions can facilitate healing.[2]

Systemic treatment is indicated when more severe or widely spread lesions are present. Treatment for Hurley Stage II and resistant Hurley Stage I involves oral antibiotics.[5] Acute abscesses may be treated with flucloxacillin 500mgs qds for 7 days. Tetracyclines such as Tetralysal 300mgs once daily or Minocycline 100mgs once daily may be considered. The combined use of systemic clindamycin and systemic rifampicin (Rifampicin 300mgs twice daily and Clindamycin 300mgs twice daily for 10 weeks) under specialist supervision has proven beneficial, with variable results.[9]

For Hurley stage III and resistant lower stages, tumour necrosis factor-alpha inhibitors are indicated. Recent studies have shown that biologics, adalimumab and infliximab, two different monoclonal antibodies against TNF-α, are effective in the treatment of moderate to severe HS (Hurley II–III), with improvement in the patient's quality of life, with adalimumab being more tolerable.[5, 7] Adalimumab (Humira) is licensed in Ireland for the treatment of severe hidradenitis suppurativa, 160mgs Day 1, 80mgs day 15 and 40mgs day 29, followed by weekly injections of 40mgs subcutaneously. [9]

Surgery at later stage HS is often required, and involves a wide excision to include the lesions, tracts, and scars of an entire affected area. A combination of medical treatment and surgical excision is often the preferred approach. Other therapeutic options may include localised laser and pulsed light therapy which can help to disrupt the inflammatory process.[2]

Pain management is important. The pain of HS is both inflammatory and non-inflammatory. Sources of pain can include scarring, keloids, abscesses, open ulcerations, sinus tracts, frictional pain, lymphedema, anal fissures, and arthritis. Depending on disease severity and type of pain, topical agents such as lidocaine and anti-inflammatories, systemic nonsteroidal anti-inflammatories, acetaminophen, atypical anticonvulsants including gabapentin or pregabalin, and serotonin-norepinephrine reuptake inhibitors may be beneficial.[2]

Treatment should include management of comorbidities that contribute to the development or worsening of the disease process. Individuals who are overweight or who smoke usually have more severe disease progression, therefore, help with weight loss and smoking cessation are important components of treatment. Avoidance of tight and synthetic clothing, harsh body cleaning products and adhesive dressings can be beneficial for patients. Patient assurance that the condition is not contagious or the result of poor hygiene can be helpful. Counselling and support groups are often helpful additions to treatment plans. [2]

Prognosis and Outlook

There is no cure for hidradenitis suppurativa and prognosis is widely variable. Outcomes can worsen if there is a delay in diagnosis and treatment during the early stages of the condition, and also if comorbid conditions of smoking and obesity are not addressed and improved. The best prognosis involves early recognition and aggressive treatment at early stages of the condition and psychosocial support.[2] HS is under-recognised and diagnosis is frequently delayed. Patients often attend their GP or local A&E to have individual abscesses incised and drained, therefore the recurring patterns is not noticed at the time and often mistaken for a simple infection.[8] As the condition often goes undetected and untreated for long periods, it is important for primary care providers to recognise hidradenitis suppurativa based on the morphology, location and chronicity of lesions, and begin the correct treatment early in the disease process. An inter-professional approach is most effective in the management of the condition. HS can cause significant psychosocial distress for patients and mental health supports from psychiatrists, psychologists, or counsellors should be provided as necessary. Therapeutic options for hidradenitis suppurativa is gradually expanding, however, further research studies are necessary to ascertain whether certain genetic, clinical, or phenotypic factors may predict or guide more effective treatments and outcomes.

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