

# Hidradenitis Suppurativa

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**CASE SUMMARY:** A 66-year-old obese (BMI 34) diabetic man, a former smoker who had quit 4 months ago, had a 20-year history of perianal, inguinal, and gluteal hidradenitis suppurativa. He presented to the office with persistent perianal and gluteal inflammation and drainage. He had had various surgical excisions of the affected area over the preceding 10 years. The patient was also following up with dermatology and was previously on doxycycline and infliximab with little improvement in his symptoms. On physical examination of his gluteal and perianal region, he had multiple areas of scarring and draining sinus tracts with significant induration (Figs. 1 and 2). The patient underwent extensive unroofing of the sinus tracts and excision of active disease (Fig. 3).

## CLINICAL QUESTIONS

- What is the pathogenesis of hidradenitis suppurativa?
- What other diseases should be considered in a differential diagnosis that includes hidradenitis suppurativa?
- How is hidradenitis suppurativa staged and managed?

## BACKGROUND

Hidradenitis suppurativa (HS), or acne inversa, is a chronic and recurrent inflammatory disease of the skin and subcutaneous tissue most commonly affecting the axillary,

inframammary, groin, perineal, and perianal regions. The disease process has been thought to result primarily from a chronic inflammatory condition of the apocrine sweat glands.<sup>1-4</sup> The current theory is that HS is caused by a chronic follicular occlusion and rupture, leading to secondary inflammation of surrounding tissue, including apocrine sweat glands.<sup>1,5</sup> Another complementary theory is that HS relates to an autoinflammatory process.<sup>6</sup>

Hidradenitis suppurativa is most often seen in women in the second or third decades of life,<sup>1,5</sup> yet the perianal subtype has been described as a more common occurrence in men and in the third and fourth decades.<sup>2,5</sup> Previous French and Danish studies have noted a prevalence rate between 1% and 4% in their populations, while a large health insured-only American study found a prevalence of 0.053%.<sup>1,6</sup>

Follicular occlusion refers to dysfunction of the follicular pilosebaceous unit, which leads to follicular rupture, secondary inflammation, and possible bacterial infection



**FIGURE 1.** Gluteal hidradenitis suppurativa.

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**FIGURE 2.** Previous scarring from perianal hidradenitis suppurativa excision.

of the apocrine glands.<sup>1</sup> This process can clinically manifest as chronic superficial sinus tract formation, scarring, and abscesses and also has been described in other disorders. The follicular occlusion tetrad includes HS, acne conglobata (severe cystic acne), dissecting cellulitis of the scalp, and pilonidal sinus.<sup>1,6</sup>

The connection between HS and IBD may point to an autoinflammatory etiology. Pyoderma gangrenosum (PG), a manifestation of ulcerative colitis, is a painful, inflammatory skin disorder. Pyoderma gangrenosum and HS have been linked in 3 reported syndromes: PASH (PG, HS, and acne conglobata), PASS (PG, HS, acne conglobata, and axial spondyloarthritis), and PAPASH (PG, HS,

acne conglobata, and pyogenic arthritis).<sup>6</sup> PASH and PASH share a common missense mutation that leads to an increase in neutrophil-mediated inflammation.<sup>6</sup> These syndromes help further support a possible autoimmune link for HS.

Hidradenitis suppurativa has been associated with other comorbidities and lifestyle risk factors including obesity, smoking, diabetes mellitus, and hyperlipidemia.<sup>1,3</sup> One study found that 70% of patients with perianal HS were smokers.<sup>3</sup> These documented associations are not the primary cause of the disease process, but, rather, they are all possible exacerbating factors.<sup>1</sup> Crohn's disease has also been linked to HS. A small case series from the Cleveland Clinic colorectal group found that over one-third of their patients with HS had a diagnosis of Crohn's disease, and the diagnosis of Crohn's disease was made on average 3.5 years before the diagnosis of HS.<sup>7</sup>

The Hurley staging system for HS has been used to help classify the severity of the disease for individual patients. This very simple classification system does not address the overall disease burden on the patient, or the extent of surgical operation that may be needed. Stage I is defined as one or more abscesses without any sinus tract formation. Stage II is single or multiple abscesses with sinus tracts and scarring. Stage III refers to multiple interconnected tracts and abscesses in one anatomic area.<sup>1,5</sup>

## PRESENTATION AND DIAGNOSIS

Hidradenitis suppurativa most commonly presents in the axilla, and up to one-third of patients with HS have perianal or perineal involvement.<sup>1</sup> Symptomatic patients typically present with painful, deep nodules that may be associated with abscess and/or sinus tract formation, depending on the stage of presentation.<sup>1,5</sup> Hidradenitis suppurativa in the anal canal is superficial to the internal anal sphincter, is distal to the dentate line, and has no relation to the intersphincteric space or cryptoglandular units.<sup>4</sup> A lesion observed proximal to the dentate line should raise suspicion for Crohn's disease, although the 2 disease processes can exist simultaneously.<sup>3,4</sup> Fistulas can develop in this area with chronic, poorly controlled perianal HS.<sup>3</sup>

The diagnosis of HS is clinical. Laboratory and/or imaging tests are not necessary for the diagnosis. The key characteristics on the physical examination are sinus tract and scar formation. Pathological examination of HS specimens can demonstrate follicular plugging accompanied by fibroplasia and a mixed inflammatory cell infiltrate.<sup>1</sup> Differential diagnoses for perianal HS include fistula-in-ano, Crohn's disease, furunculosis, and pilonidal sinus. Fistula-in-ano, unlike HS, can involve the intersphincteric plane and be proximal to the dentate line.<sup>4</sup> Perianal Crohn's di-



**FIGURE 3.** Postexcision and unroofing of gluteal hidradenitis suppurativa.

sease is usually accompanied by other colorectal manifestations. A furuncle has a necrotic plug, which is not seen with HS abscesses. A midline pit overlying the coccyx is characteristic of pilonidal sinus, a common standalone lesion that can also be, along with HS, part of the follicular occlusion tetrad.<sup>4,6</sup>

## MANAGEMENT

The management of HS remains controversial. There are differing strategies from discipline to discipline. Dermatologists tend to take a more stepwise medical approach compared to surgeons. Nonetheless, there is no single best modality to treat every patient with HS. Assessing the Hurley stage may be helpful in traversing therapeutic strategy. With respect to perianal HS, a thorough history and physical examination can also help discern the level of debilitation. Perianal HS can significantly affect a patient's quality of life. The traditional approach to HS has been conservative medical therapy before gradually increasing levels of surgical treatment. With the perianal HS population, a delay in definitive diagnosis is typically encountered, and, therefore, a more aggressive treatment tactic may be warranted.<sup>8</sup>

Several aspects of the daily life of a patient with HS can be optimized as part of the initial treatment. When appropriate, recommendations for weight loss, smoking cessation, and glycemic control can be helpful conservative measures.<sup>8</sup> A reasonable first-line medical treatment for mild to moderate HS is topical clindamycin 1% or oral tetracycline.<sup>1,8</sup> Refractory cases are typically started on a combination of oral clindamycin and rifampicin, which has shown an 82% treatment response and 47% total remission rate.<sup>9</sup> Overall, topical and/or oral antibiotics do not offer a complete and lasting resolution for HS.

Tumor necrosis factor inhibitors have demonstrated good results for refractory HS cases. Randomized controlled studies (PIONEER I and II) looked specifically at adalimumab in patients with moderate to severe HS. A significant treatment response with the use of adalimumab as

a monotherapy was observed.<sup>10</sup> Infliximab, although not Food and Drug Administration-approved for HS, has also been studied and similarly showed improvements in patients with Hurley II and III stage HS.<sup>1,8</sup>

The surgical treatment of HS varies, especially based on severity. Although there is no interruption of disease progression, an incision and drainage can be helpful for the relief of painful abscess cavities related to HS. Virtually all patients who undergo an incision and drainage for HS have a recurrence.<sup>8</sup> Unroofing, or exteriorization, of sinus tracts has been suggested as a reasonable treatment option.<sup>1,4</sup> In patients who undergo surgical excision, the recurrence rates differ based on anatomic site. Low recurrence rates have been seen with axillary and perianal regions and higher rates with submammary and inguino-perineal regions.<sup>8</sup> A series of 30 anal canal HS cases from the Mayo Clinic by Culp<sup>4</sup> found no recurrences at or near the operative sites after surgical excision with a range of follow-up from 1 to 7 years.

Several authors have compared the method of wound closure after excision as well. A meta-analysis by Mehdizadeh et al<sup>11</sup> reviewed recurrence rates of HS after surgical treatment. With a range of follow-up between 6 months and 8 years, the authors found average recurrence rates of 15% with primary closure, 8% with flaps, and 6% with skin grafting. The closure technique should be based on the functional need of the excision area to achieve optimal results.<sup>8</sup> The role of diverting colostomy in HS has not been found necessary even in very extensive disease.<sup>4</sup> Of note, surgical management of HS alone cannot address the underlying etiology of the disease.

The combination of medical and surgical therapy has shown promising outcomes in the HS population. Specifically, the addition of biologics with excision has been studied. Defazio et al<sup>12</sup> compared patients who have HS treated with surgery alone versus those treated with surgery and biologic therapy. There was a significant decrease in the rate of recurrence and disease progression in the combined group with no increased rate of complications. Ultimately, the management of perianal HS must be personalized to each patient.