Hidradenitis Suppurativa of the Anorectal Region

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ETIOLOGY
Hidradenitis suppurativa (HS) was originally described by Velpeau in 1832 (1) and named by Verneuil (2) in 1864. In 1942 Jackman (3) made the distinction between chronic hidradenitis suppurativa and anal fistula. The etiology of HS is unknown. The estimated prevalence is 4% (4). Many cases of HS have a mild course, but disease recurrence is frequent. Most cases occur between the ages of 16 and 40 years. There are conflicting reports about the predominance of the disease in men (5) and women (6). Perineal HS appears to be male-dominated in most large series (7,8). The disease is occasionally familial (9) and an autosomal dominant mode of transmission with a high penetrance has been suggested (10). The disease is associated with acne, poor skin hygiene, and hyperhidrosis. An androgen-based endocrine disorder has been postulated (11). Associations with obesity, hypercholesterolemia, and Cushing's disease have been described. Cigarette smoking has been implicated as a triggering factor (12). In patients with long-standing disease, a 3.2% incidence of squamous cell cancer has been noted (13).

Traditionally, HS has been classified as an inflammation of the apocrine gland. Many authors believe that HS begins with a keratinous plugging of the apocrine duct, followed by duct and gland dilatation and inflammation. Gland rupture spreads the infection to surrounding tissue, causing localized abscesses, chronic draining sinuses, and finally scarring and fibrosis (13,14). Others, however, suggest that the apocrine gland is secondarily infected (15). In a prospective study evaluating surgical specimens from patients with HS, a heterogeneous histologic picture was seen. Apocrine glands were involved in a minority of the 60 specimens reviewed. The disease appeared to be predominantly follicular, with poral occlusion or epithelial cyst accounting for 67% of all lesions. Apocrine gland involvement was noted in a minority of axillary lesions (<5%). A current theory for HS is that it is an inflammatory disease originating from the hair follicle. Follicular hyperkeratosis leads to retention of follicular products, with secondary infection and inflammation of apocrine and eccrine sweat glands (Fig. 1) (6).

**Fig. 1** FIG. 1. Hair follicle with acute inflammation and abscess formation.

**PRESENTATION**

The clinical appearance of HS may consist of a single nodule or multiple, discrete nodules located in the cutaneous layer and extending into the subcutaneous tissue. Lesions begin as blind boils most commonly at or soon after puberty. In contrast to staphylococcal furunculosis, these abscesses tend to be deep and rounded, with no pointing or central necrosis. The overlying skin is rarely erythematous, and enlarged pores can be seen in the indurated area. In more advanced conditions, these discrete lesions can become confluent and present as a larger area of induration and mild tenderness, and burrowing sinuses may be seen that may extend anteriorly into the groins or posteriorly into the buttocks or sacrococcygeal area (16). These sinuses may involve the posterior scrotum, labia, or proximal posterior or medial thigh. Pitted scars are from fibrotic skin contraction and the loss of one or more apocrine units (Fig. 2).

**Fig. 2** FIG. 2. Extensive perianal induration, scarring, and superficial fistula tract.

Perianal HS can mimic several common anal conditions. The differential diagnosis includes perirectal abscess and fistula, pilonidal disease, Crohn's disease, actinomycosis, and tuberculosis, lymphogranuloma venereum, and granuloma inguinale. Misdiagnosis is common; frequently, the etiology of the draining perianal sinus cannot be determined until examination under anesthesia is performed. Most of the patients from the Lahey Clinic (8) experience have been treated for other conditions before the diagnosis of HS was made.

One of the most important points of distinction between a fistulous tract originating from cryptoglandular tissue and the tract of HS is its relationship to the sphincter mechanism. The tract of HS arises within the skin of the anal canal, lies superficial to the internal anal sphincter, and has no relationship to the intermuscular space or the glands and involves the most distal area of the anus, superficial to the external sphincter. We frequently use endorectal ultrasound to identify an internal rectal opening of a fistula. Unlike other septic or fistulizing processes around the anus, HS infrequently connects with the anal canal or rectum. The anal fistula traverses deep to the internal sphincter in the intramuscular space from the origin in the cryptoglandular area and penetrates the perianal skin secondarily (7).
Differentiation of HS from Crohn's disease may be difficult, and the two diseases can coexist. Church et al. (17) described a 38% dual pathology of Crohn's proctocolitis and HS. Interestingly, the diagnosis of Crohn's disease predated HS by 3.5 years. The presence of bilateral chronic draining sinuses and tracks that traverse the midline suggest Crohn's. In Crohn's disease, the fistulization process is frequently deep to the internal sphincter, having penetrated the rectum from an ulcer in the rectal wall. Differentiating the various causes of draining perianal sinuses is of prognostic significance. When the primary source of a fistulous abscess is located in a cryptoglandular unit, recurrence is unlikely if surgical treatment is adequate. In patients with HS, new lesions may develop in the anogenital region and other apocrine-bearing regions.

MEDICAL THERAPY

Medical treatment options for HS include antibiotics, isotretinoin, and antiandrogen agents. Antibiotics are helpful in diminishing the inflammatory response to the sebum in the acute phase. Patients occasionally derive symptomatic benefit from long-term antibiotic therapy, but relapse is inevitable when treatment is withdrawn. In the early stages pus is sterile, but with disease progression bacteria superinfect the lesions. Often there is only a small quantity of serous material associated with extensive pitting induration and sinuses. Only topical clindamycin has been shown to be effective in a double-blind placebo-controlled study (18). Antibiotics have no effect on chronic HS. The primary pathogen cultured from patients with HS is Streptococcus milleri. Other organisms identified include Staphylococcus aureus, Escherichia coli, Proteus, anaerobic streptococcus and Bacteroides, and Chlamydia trachomatis (19) species.

Isotretinoin (Accutane) is a synthetic vitamin A derivative for the treatment of severe nodulocystic acne. The mechanism in HS is not completely understood, but the antiinflammatory effects are thought to be a major component. Isotretinoin has been shown to be useful, but randomized prospective studies are needed to determine its role (20) in this condition.

Treatment with antiandrogen cyproterone acetate in combination with estrogen has been shown to be of some benefit (21). A report of two patients showed improvement with finasteride (22), a competitive inhibitor of the 5-alpha-reductase type 2 isoenzyme; its main side effect was impaired androgenization.

SURGICAL THERAPY

Patients whose condition is refractory to medical therapy or for those who have chronic long-standing HS, surgical therapy is the only effective option. Surgery ranges from drainage to radical disfiguring excision. There is no standard treatment, and most studies lack long-term follow-up. Incision and drainage of individual lesions and abscesses may provide temporary relief of symptoms, but this does not cure the underlying sinuses and infected apocrine glands, and recurrence rates are as high as 100% at 3 months (23). Most authors agree that wide excision is the key to preventing recurrence of perianal HS, and recurrence rates range from 0% to 60% (7,8,22).

The method of wound closure may also have an effect on recurrence. Excision with primary closure, excision with packing, excision with grafting, and excision with marsupialization are four accepted surgical methods. In some cases all four methods may be applied to the same patient. Primary wound closure requires a somewhat limited resection and has been associated with a higher recurrence rate (22). This is the preferred method of Corman et al. (24) in selected patients with small lesions. Wide excision with healing by second intention is the most common method of treatment. The Mayo Clinic experience (132 patients with HS, 31 with perianal HS) quoted a recurrence rate of 0% after wide excision with exposure of the entire lesion and preservation of the floor of the track for epithelial regenerative elements (8). This method provides good results, with epithelization in 6 to 8 weeks.

The experience with skin grafting is variable. The Lahey Clinic series (8) shows a uniform failure with skin grafting, but other series have better results (25). A diverting colostomy is rarely necessary unless extensive surgery and grafting is planned. Usually colostomy can be avoided because HS does not involve the sphincter mechanism and continence is maintained. If stool contamination will prohibit healing of the wound, a temporary diverting colostomy is a good option.
Recurrence rates after excision for HS are associated with disease site. Axillary and perianal HS frequently has a mild course, whereas inguinal HS is often associated with a high recurrence rate and debilitating outcome (22).

**SUMMARY**

Surgical therapy remains the mainstay of treatment for HS. There have been few advances for definitive treatment and cure over the centuries. It is now thought that HS is an inflammatory process of the hair follicle and despite its name is not initiated by infection and does not primarily affect the sweat glands. The key to a successful outcome in a patient with HS is making the correct diagnosis, performing adequate surgical excision, and continuing with long-term follow-up. Smoking cessation and cancer surveillance should be a routine part of treatment protocols.

**REFERENCES**


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