Hidradenitis Suppurativa

Hidradenitis Suppurativa is a chronic, inflammatory scarring disease of the apocrine sweat gland-bearing skin. The disease is characterized by the presence of multiple abscesses, fibrosis, and sinus tracts. A number of synonyms are used to indicate hidradenitis suppurativa such as apocrinitis, hidradenitis axillaris, and abscess of the apocrine sweat gland. Treatment is usually very difficult.

Hidradenitis suppurativa was first described by Velpeau in 1839. Verneuil suggested that the etiology was abscess formation of sweat glands. 19 Histologic features of hidradenitis suppurativa were first described in 1939. 19

Epidemiology

The disease occurs in both sexes, usually in the second or third decade, and affects women more than men. The disease is rare before puberty. No racial predilection is described. The prevalence of hidradenitis suppurativa is estimated at 4% of women in the general population. 20 The prevalence was 1% in the general Danish population and 4 percent in a selected group of young adults attending a sexually transmitted diseases clinic. 21 The different age groups examined and differences in diagnostic specificities may account for the variable prevalence estimates.

Etiology and Pathogenesis

The pathogenesis of hidradenitis suppurativa is unknown. The anatomic location of the disease in the axilla, groin and perianal and inframammary suggests a disorder of apocrine glands. It has been suggested that occlusion of the apocrine duct may lead to severe dilatation and apocrine gland inflammation, with ensuing bacterial growth and neutrophilic inflammation of the duct and surrounding tissue, fibrosis, and sinus tract formation. 22

The clinical similarity of hidradenitis suppurativa to and its frequent association with the follicular occlusion-related diseases, such as acne conglobata, and with dissecting cellulitis of the scalp suggest that hidradenitis suppurativa may share the same pathogenesis. Follicular occlusion is a prominent feature in all biopsy specimens from patients with hidradenitis suppurativa. 23-25 At a later stage, apocrine glands become inflamed due to the direct continuity of the apocrine duct with the follicular infundibulum. The findings of primary folliculitis and perifollicular inflammation with normal eccrine and apocrine glands in biopsy specimens support the hypothesis that hidradenitis suppurativa is a follicular disease and that the apocrine gland inflammation may be secondary or incidental. Apoeccrine glands, whose secretory coils show decapitation secretion with a straight intradermal duct opening directly onto the surface of the skin, remain intact until dermal fibrosis distorts the opening duct, causing irritation by the apocrine sweat. 21

The role of androgens or other hormones in hidradenitis suppurativa is unclear. Obesity, a common finding in hidradenitis suppurativa, is known to alter sex hormone metabolism, leading to an androgen-excess state. Excess androgens can lead to coarsening of the hair shaft and hence to follicular occlusion. 22 Women with hidradenitis suppurativa tend to have a shorter menstrual cycle and a longer duration of menstrual flow, suggesting a possible hormonal influence. 20 However, the incidence of acne, hirsutism, and irregular menstrual periods–signs of a hyperandrogenic state–is not different from that in the general population. There is no difference in the plasma levels of androgens between hidradenitis suppurativa patients and controls. 26 However, cyclic variations of the hormonal cycle with the amelioration of the disease during the estrogen-elevation phase and flares during the estrogen-fall phase, have been reported. 26 Furthermore, improvement is noted during pregnancy and flares may be seen post partum. A genetic factor may be important in the etiology of the disease, and a positive family history of hidradenitis is often present. 20

Clinical Manifestations

patients with hidradenitis suppurativa are usually otherwise healthy. Obesity is common, as are follicular occlusive diseases, such as acne conglobata, and dissecting cellulitis of the scalp. the early lesion is a tender, red dermal abscess measuring about 0.5 to 2 cm. The lesion may resemble an inflamed epidermoid cyst. The abscess gradually increases in size and may open to the surface, discharging purulent or seropurulent material, if untreated. the inflammation subsides gradually; however recurrent episodes of inflammation in the same abscess or others is the rule. Inflammatory abscesses may appear at other anatomic areas at the same time. Fibrosis eventually becomes prominent, and healing appears incomplete (Fig. 77-4). In advanced stages, bands of scar tissue and bridging fibrosis develop that may restrict the mobility of the tissue (Fig. 77-5). Three clinical stages of hidradenitis suppurativa were described by Hurley 27 and are summarized in table 77-1.

The axilla, buttock, inguinal, perianal, mammary and inframammary areas are the most commonly affected. The abdominal wall, chest, scalp, and lower extremities may also be involved. Although apocrine sweating is absent in chronic cases, the lesions have a foul smell secondary to bacterial overgrowth and colonization. Sinus tracts are often seen, draining minimal purulent discharge. Perianal hidradenitis can extend to involve the anus and the rectum. Sinus tract formation and anal canal fibrosis
can lead to stricture formation. Urethral and vaginal fistulas may develop with deep vaginal involvement. mild arthritis of peripheral and axial type may be seen in acute flares. Laboratory evaluation reveals occasional elevation of white blood cells. *Staphylococcus, Streptococcus,* and *Escherichia coli* are the organisms most commonly cultured from the draining material. *Proteus, Pseudomonas,* and anaerobes have been encountered occasionally.

### Pathology

The histologic findings in hidradenitis suppurativa cover a wide spectrum of changes. Follicular plugging and poral occlusion are prominent features. Occasionally, large cystic spaces are seen, filled with keratin material and hair shafts and lined by stratified squamous epithelium resembling epithelial cysts. Fibrosis is often seen in the dermis and may occasionally extend to the subcutaneous tissue. Various degrees of inflammation and fibrosis are seen, according to the stage of the disease. Folliculitis and perifollicular inflammation are common and seen in about two-thirds of cases, with or without follicular occlusion. The inflammatory cells are composed of neutrophils, lymphocytes, plasma cells and occasional eosinophils. Areas of frank dermal abscess (Fig. 77-6). Active inflammation around sweat glands is less common than that around hair follicles. Apocrine gland destruction by neutrophilic infiltrate is seen occasionally in the axilla. Apocrinitis, originally suggested as the typical histologic appearance of hidradenitis suppurativa, is seen in a minority of cases. (Fig. 77-7). Foreign body type granulomas around hair follicles and sinus tracts are occasionally seen. The presence of epithelioid granulomas in the dermis away from the areas of inflammation is a rare finding (5 percent) and should alert the clinician to the possibility of coexisting Crohn's disease or sarcoidosis.

(Fig. 77-4 omitted: Multiple abscesses in the axilla of a patient with stage 1 hidradenitis suppurativa. No fibrosis is yet present in the involved area.)

(Fig. 77-5 omitted: Multiple interconnected sinus tracts with severe fibrosis representing end-stage hidradenitis suppurativa. A new abscess is present at the edge of the axilla.)

(Fig. 77-6 omitted: Dermal abscess with dense inflammatory cells composed of neutrophils, lymphocytes and a few plasma cells. Note the intense perifollicular inflammation.)

(Fig. 77-7 omitted: Apocrine glands filled with a neutrophilic infiltrate, giving the appearance of “apocrinitis.”)

<table>
<thead>
<tr>
<th>Clinical Staging of Hidradenitis Suppurativa</th>
<th>Stage I</th>
<th>Solitary or multiple isolated abscess formation without scarring or sinus tracts</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage II</td>
<td>Recurrent abscesses, single or multiple widely separated lesions, with sinus tract formation and cicatrization</td>
<td></td>
</tr>
<tr>
<td>Stage III</td>
<td>Diffuse or broad involvement across a regional area with multiple interconnected sinus tracts and abscesses.</td>
<td></td>
</tr>
</tbody>
</table>

**TABLE 77-1**

**Diagnosis and Differential Diagnosis**

Any inflammatory, abscess-like swelling of the apocrine gland-bearing skin should be regarded as possible hidradenitis suppurativa. A solitary abscess in the early stages resembles a carbuncle, lymphadenitis, or an infected epidermoid cyst. In the vaginal area it resembles an infected Bartholin cyst. Multiple abscesses in different apocrine gland-bearing skin areas should present no difficulty in establishing the diagnosis. The distribution and characteristic scarring, sinus tracts and partial healing of infected abscesses are usually diagnostic. Other dermatoses that produce fistulas should be included in the differential diagnosis, such as tuberculosis, actinomycosis, tularemia, and cat-scratch disease. In the inguinal area, lymphogranuloma venereum, granuloma inguinale, Crohn's disease, and ulcerative colitis should be excluded.

**Treatment**

Treatment of hidradenitis suppurativa is very difficult. medical management is recommended in early stages; however, a surgical approach is preferred if scarring and sinus tract formation have developed.

For early disease, intralosional triamcinolone acetonide (5 to 10 mg/mL) may help. Incision and drainage are helpful if spontaneous rupture is imminent. In these situations, drainage and intralosional glucocorticoid injections can be done simultaneously. Bacterial culture of the drained material should be done, and antibiotics should be selected based on sensitivity. concomitant use of antibiotics is recommended. Minocycline, ciprofloxacin, cephalosporins, clindamycin, or semisynthetic penicillins can be used in the usual doses for soft tissue infections. Isotretinoin may be helpful in some patients. The dose is 1mg/kg, as in acne patients; however higher dosages or longer duration may be needed. The response rate is less than 50 percent. Once significant fibrosis ensues, retinoids are less effective. If inflammation is severe, a short course of systemic glucocorticoids (prednisone, 40 to 60 mg, to be tapered over 2 to 3 weeks) is often needed.

Sawers et al. reported successful control of the condition by a regimen of 100mg of cyproterone acetate from days 5 to 14 and 50ug of ethinylestradiol from days 5 to 25 of each menstrual cycle. In a double-blind trial, a more moderate but significant control was achieved by using 50mg of cyproterone acetate. Cyclosporine has also been used successfully. Local care includes gentle cleansing with antiseptic soaps or cleansers and application of warm compresses with saline or Burrow's solution. Some have advocated the use of 6.25% aluminum chloride hexahydrate in absolute ethanol as an antiperspirant and antibacterial agent. Also suggested are avoidance of tight fitting clothing and roll-on antiperspirant-deodorants, which can produce frictional trauma and exacerbation of the disease.

If scarring develops in conjunction with inflammation, exteriorization is preferred. The aim of the procedure is to destroy the sinus tracts and drain the abscessed area. Fibrotic tracts are explored by soft probes, and complete destruction of granulation tissue and epithelial cells is attempted, even from the smallest
pockets. Curettage and electrofulguration of the base are used to destroy residual infected tissue. The wound is then covered by sterile gauze and cleaned twice daily with warm compresses and topical antibiotic ointments. The use of oral antibiotics is recommended on an individual basis. The wound usually heals from 4 to 8 weeks, and the scar is usually smooth and acceptable. This procedure can be performed in all locations except the perianal area, where the risk of traumatizing the anal sphincter is high. Simple excision and primary closure can be done for smaller lesions, but recurrences are more likely.

In stage III disease, complete removal of the affected area is often required. Wide local excision and healing by secondary intention is considered the surgical treatment of choice. Large wounds in the genital area are best covered with synthetic dressings, with healing by secondary intention. Surgical excision using a CO2 laser and second-intention healing is often associated with good results and minimal complications. Healing usually occurs in 4 to 8 weeks. One promising treatment is CO2 laser ablation of lesions – by vaporizing the tissue in layers until all the macroscopically abnormal tissue has been removed. Sparing of normal tissue is achieved using this method, while surgical removal of a lesion by scalpel or laser may remove healthy tissue unnecessarily.

**Course and Prognosis**

Hidradenitis suppurativa is a chronic disease that progresses relentlessly despite treatment, but the early stages may be curable with proper diagnosis and treatment. The psychological impact of the disease exceeds the systemic complications. Patients with advanced disease are often depressed and socially isolated. Secondary infection of the lesions, which causes a foul smelling odor, adds to the misery of some patients. Septicemia is a rare event. Restricted mobility, secondary to extensive scarring, and fistula formation are common complications. Urethral and rectal fistulas and strictures are uncommon, and squamous cell carcinoma is a rare complication of long-standing disease. The majority of cases develop on the buttocks and perianal areas. Squamous cell carcinoma arising from hidradenitis suppurativa usually behaves aggressively with local invasion, distant metastasis, and high mortality. Other rare complications include amyloidosis, interstitial keratitis, and anemia.

**REFERENCES**


