Perianal Hidradenitis Suppurativa*

A Clinical and Pathologic Study

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Hidradenitis suppurativa is a chronic inflammatory condition affecting the skin and subcutaneous tissues in those regions in which apocrine sweat glands are said to be found, namely the axilla, about the areola of the breast, in the inguinal regions, about the external genitalia and in the perianal region. Late in its course, the disease is characterized by the formation of subcutaneous abscesses, fistulas and sinuses, and by cutaneous scarring.

As is true in most conditions that neither represent a great threat to life nor afflict large numbers of people, this disease has not attracted widespread attention among physicians. Most of the reports concerning hidradenitis represent impressions based on a relatively narrow clinical experience. It is the purpose of this paper to present the results of a clinical and pathologic study of a sizeable group of cases of hidradenitis, comparing our findings with comparable data gleaned from the literature.

General Background

Velpeau made the first contribution to the literature on the subject of hidradenitis suppurativa when, in 1839, he described the clinical entity consisting of circumscribed inflammation with superficial abscesses in the axilla, about the anus and at the nipple. Krause and Robin, in 1845, observed that the sweat glands in the axilla and inguinal regions were different from those in other parts of the body. From 1854 to 1865, Verneuil wrote many papers concerning hidradenitis and apparently was the first to suggest a relationship between this disease and the apocrine sweat glands.

American contributions on the subject commenced with Cole and Driver, who reported a case of hidradenitis in 1929. In 1933, Lane contributed a comprehensive review. In 1937, Brunsting combined a critical survey of the literature with data in 22 cases that he had studied personally.

The concept that hidradenitis suppurativa is related in some way to the apocrine sweat glands appears to be universally accepted in the literature, apparently stemming from the fact that the disease is found most often in those regions where apocrine glands are said to be.

Anatomic Considerations: In 1887, Ranvier classified the cutaneous glands on the basis of the mechanism of secretion, dividing them into holocrine (sebaceous) and merocrine (sweat) glands. In 1921, Schiefferdecker subclassified the latter into eccrine and apocrine glands. The long-accepted distinguishing characteristics of apocrine and eccrine sweat glands are summarized in the accompanying table.
Table 1. Distinguishing Characteristics of Eccrine and Apocrine Sweat Glands

<table>
<thead>
<tr>
<th></th>
<th>Eccrine Glands</th>
<th>Apocrine Glands</th>
</tr>
</thead>
<tbody>
<tr>
<td>Origin</td>
<td>From epidermis</td>
<td>From anlage of hair follicle</td>
</tr>
<tr>
<td>Time of activity</td>
<td>From birth</td>
<td>Begins at puberty</td>
</tr>
<tr>
<td>Distribution</td>
<td>Diffuse, with few exceptions</td>
<td>Axilla, groin, genitalia, breast, perianal region</td>
</tr>
<tr>
<td>Type of gland</td>
<td>Simple tubular</td>
<td>Large compound tubular</td>
</tr>
<tr>
<td>Material secreted</td>
<td>Watery</td>
<td>Thick and milky</td>
</tr>
<tr>
<td>Mode of secretion</td>
<td>Protoplasm not lost from cells of gland</td>
<td>Protoplasm lost from cells of gland</td>
</tr>
<tr>
<td>Site of duct</td>
<td>On epidermis independent of hair follicle</td>
<td>Opens into or adjacent to hair follicle</td>
</tr>
<tr>
<td>Color</td>
<td>Yellow</td>
<td>Reddish</td>
</tr>
<tr>
<td>Size</td>
<td>Small</td>
<td>Large</td>
</tr>
<tr>
<td>Myo-epithelial investment</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Staining with hematoxylin and eosin</td>
<td>Basophilic</td>
<td>Acidophilic</td>
</tr>
<tr>
<td>Iron stain</td>
<td>Negative</td>
<td>Positive in 70 per cent</td>
</tr>
</tbody>
</table>

The function of the apocrine glands apparently is regulated to some extent by hormonal control. Most investigators agree that the apocrine glands probably do not become physiologically active until puberty. It has been suggested that these glands in man are a vestigial remnant of the scent glands of sexual function in the lower animals. Some studies indicate that they enlarge and secrete more actively just before and during menstruation. Both an increase and a decrease in activity of these glands during pregnancy have been reported. Physiologic and histologic atrophy of the glands apparently follows the menopause.

Etiology of Hidradenitis Suppurativa: Many factors that might contribute to hidradenitis have been mentioned. These include uncleanliness, cold, friction, irritating applications, dyspepsia, constipation, hemorrhoids, pruritus, cachexia, depilatories or shaving, anhidrotics and moisture due to apposition of two cutaneous surfaces. Closure of the pores has been suggested as a contributing factor, but Hurley and Shelley demonstrated experimentally that such closure does not produce disease of the apocrine glands. Most investigators agree that specific organisms have no significance in the production of hidradenitis.

Pathogenesis: Nearly all writers agree that the infection in hidradenitis is exogenous, but some disagreement exists whether the apocrine glands are involved primarily or secondarily. Some investigators consider that the causative agent enters through the hair follicle and the excretory duct of the apocrine gland, whereas others contend that the apocrine glands are involved only by contiguity.

Signs and Symptoms: The earliest symptoms are usually insidious in onset and consist of burning, pruritus, local heat, hyperhidrosis and, eventually, pain. As the inflammatory process increases, firm subcutaneous nodules may be noted. The process either may undergo involution at this point or progress further, with adjacent inflammatory nodules coalescing to form cordlike bands. The skin in the involved region frequently becomes either red or bluish red. A small amount of watery pus may drain
from the lesion. With or without such drainage, gradual resolution of the entire process may ensue or a series of recurrences and remissions may occur.

Chronic persistent disease eventually may produce ulceration of the skin. Ulcers so produced usually have a typical appearance, with ragged, rolled-in, boggy edges and clean granulation-tissue bases. Scarring is also prominent in chronic disease, the scars usually being depressed and often producing bridges (Fig. 1). Chronic infection in the subcutaneous tissue eventuates in the formation of sinuses.

If hidradenitis of the perianal region proceeds to the point of sinus formation, these sinuses may perforate the rectum. When the anorectum is involved, the sinus usually penetrates the anus distal to the dentate line.

Systemic symptoms are typically absent in hidradenitis; if present, they are extremely mild.

**Differential Diagnosis:** Numerous conditions might be confused with hidradenitis suppurativa. Early lesions may resemble furuncles, carbuncles, lymphadenitis, cellulitis or erysipelas; however, these conditions are more acute and more frequently are accompanied by pronounced systemic reaction than is true of hidradenitis.

The late phase of hidradenitis in the perianal region might be confused with pilonidal disease, ischiorectal abscess, tuberculous abscess, noduloulcerative syphilis or anal fistula. Consideration of the history, severity of the systemic reaction, appearance and distribution of lesions, and the use of biopsy, cultures, inoculation of guinea pigs, Mantoux tests, serologic tests for syphilis and the Frei test should enable one to arrive at the correct diagnosis.

**Treatment:** With the passing of time, the utilization of certain therapeutic tools, such as autogenous vaccines, staphylococcus toxoid, injection of whole milk, and use of ultraviolet and infrared light, has been discontinued, as it has become apparent that they are not useful. The administration of thyroid extract with a low-fat diet has been suggested recently, as has the use of testosterone. However, the patients mentioned in support of these suggestions did not appear to have made unusually good recoveries. The use of antibiotics, both by parenteral administration and by local injection, has been advocated by many. Little evidence exists to attest to their efficacy in this condition.

Use of roentgen therapy in the management of hidradenitis was widespread two and three decades ago. It is still employed but its use ordinarily is restricted to cases of early and mild disease.

Surgical intervention apparently has played an increasingly important role in the treatment of hidradenitis suppurativa. Excision of the involved skin and subcutaneous tissue has come to be advocated widely for all but the mildest lesions. It is said that one should not delay employment of radical surgical treatment, since a sympathetic relationship appears to exist among the various sites involved by this
disease. One might prevent the occurrence of infection in one site or produce remission of mild lesions in such a site by surgical removal of seriously involved tissue in another location.

In many parts of the body, surgical excision of hidradenitis is followed immediately by primary closure or by the employment of split-thickness skin grafts. However, in the perianal region, it is usually best to allow the wounds to heal secondarily because they are contaminated and it is difficult to immobilize this region. Even large wounds heal rapidly by granulation in this zone.

Material and Methods

A study was made of the records of 117 patients seen at the Mayo Clinic during the decade beginning January 1, 1940, who were considered to have hidradenitis suppurativa of the perianal region. Of these 117 patients, 64 were treated surgically at the clinic. This group of 64 patients provided material for pathologic study. From the preserved gross specimens available from these 64 patients, a total of 261 sections were prepared and stained with hematoxylin and eosin.

Clinical Findings

Sex and Age: Contrary to most reports in the literature, this study showed that hidradenitis occurred almost twice as often in males as in females; 65 per cent of our patients were males. Study of the age at onset of the disease revealed that the highest incidence (26.5 per cent) was in the second decade of life; however, the incidence during the third and fourth decades was almost as great as during the second. Only 4.5 per cent of the patients noted the onset of their disease during the first decade, and 17 per cent noted its onset after the fourth decade. The oldest patient was 68 years of age when his hidradenitis began.

Sites of Involvement: From the literature, one gains the impression that certain patients have a nicely localized zone of hidradenitis lying within 3 or 4 cm. of the anal margin. Actually, one finds in studying a large series of cases that the term "perianal hidradenitis" must be used rather loosely, since many patients with disease in the anal region also have involvement of the scrotum, buttocks, proximomedial aspect of the thighs and the inguinal and pubic regions (Fig. 2). A line of demarcation usually does not exist between the anal involvement and that in the other regions.

It was evident from our studies that the perianal region was involved in about one of every six patients who had hidradenitis and that the perianal region alone was involved in one of every 13 patients who had this disease.

In this series, it was not unusual for hidradenitis to be found in regions other than those which for so long a time have been called typical. Cases were found in this study in which the posterior part of the neck, the scalp, shoulders, extremities, abdomen or face was involved synchronously or metachronously by the disease.

Symptoms and Signs: The symptoms and signs described in our cases agreed well with the classic clinical descriptions of hidradenitis suppurativa in the literature. It is indeed a chronic recurring disease.

Coexistent Diseases: In our series, a variety of unrelated anorectal pathologic conditions was mentioned, such as hemorrhoids, polyps and enlarged anal papillae. In seven cases, surgical intervention had been done for anal fistulas prior to the patients' visit to the clinic. In an eighth case, a sinus with a palpable tract leading to the anus was noted; however, the suspected communication between this sinus and the anal canal was unproved in this instance because the recommended surgical treatment was refused. Some writers have concluded that it is not unusual for the sinuses of hidradenitis to involve the anal canal, thereby producing anal fistulas. However, on the basis of this study, one must conclude that such involvement is rare.
The literature does not indicate that hidradenitis is related to pilonidal disease. In this study, a diagnosis of pilonidal disease had been made in 16 of the 117 patients. Fourteen of these 16 patients gave histories of having had surgical procedures for pilonidal disease done elsewhere. The diagnosis of pilonidal disease was made at the clinic in two instances only, and the diagnosis in one of these subsequently was changed to hidradenitis of the postsacral region.

It would appear that some common etiologic factor might be present in acne and hidradenitis. Acne had been present in at least 30 per cent of the patients in this study. The high incidence of coexistence of these two diseases has been noted by others.

**Endocrine Effect on Hidradenitis:** An endocrine effect on hidradenitis was suggested in just ten of the cases in this study. The findings in these cases were in complete agreement with what has been reported by other workers. Thus, it was noted in five cases that the disease became worse shortly before menstruation. The effect of pregnancy on hidradenitis was variable. In two cases, the onset of the disease coincided with menarche. The fact that the disease had its onset during the second, third or fourth decade in 75 per cent of the cases in this series indicates in a general way that some relationship probably exists between endocrine activity in the body and the activity of hidradenitis.

**Pathologic Findings**

**Gross Features:** The most frequently noted gross finding was that of scarring and roughening of the epidermis. Small subcutaneous sinuses, which were often interconnecting, were frequently present. Dermal and subcutaneous fibrosis was characteristic. Grossly recognizable ulceration was rare.

**Microscopic Features:** Study of the 261 sections taken from the surgical specimens in these 64 patients disclosed that eccrine glands were present in 59 of the cases, whereas typical apocrine glands were identifiable in only seven. Since some sections were taken from the least involved zones in each case, complete destruction of the apocrine glands by the inflammatory process would not appear to explain the infrequency with which apocrine glands were found.

The inflammatory cellular exudate consisted primarily of plasma cells, with some lymphocytes and fixed connective-tissue cells in nearly all cases studied, which was considered to be consistent with the chronicity of the clinical symptoms and signs. The distribution of the cellular exudate was similar in most cases. Perivascular infiltration was noted in the tissues of 42 of the 64 surgical patients. Zones of diffuse cellular inflammation were present in 39 of the patients, and giant cells were noted in these zones of diffuse inflammation in 12 instances. Cellular exudate also was noted rather frequently just beneath the epidermis, as well as in the tissue spaces or small lymphatic spaces.
Malignant change, in the form of squamous cell epithelioma, was present in two cases, in both of which eccrine sweat glands and typical inflammatory changes of hidradenitis suppurativa were noted in addition to the malignant tumor. The histories were typical of hidradenitis of long standing in both these cases, the disease having been present for 25 years in one instance and 32 years in the other (Fig. 3).
Pathogenesis of Hidradenitis

The findings in our study agree with those of Török and Talke concerning the pathogenesis of hidradenitis, indicating that the apocrine sweat glands are infected secondarily rather than primarily. Periglandular inflammatory exudate is seen frequently, but in these instances exudate usually occurs also about the eccrine glands, hair follicles, sebaceous glands and small blood vessels. It was noted that several small blood vessels usually were associated with every sweat-gland coil, whether the gland was apocrine or eccrine. It appears likely that the periglandular inflammatory exudate is actually perivascular exudate involving the small vessels about the gland coils. The periglandular inflammation could, of course, penetrate and destroy the gland from without. In no section was evidence of inflammation found within the coil of a sweat gland, eccrine or apocrine, in the absence of periglandular inflammation. On the contrary, inflammatory exudate often was noted adjacent to and among the coils of the sweat glands, while the lumen and the lining cells of these coils revealed no evidence of involvement by the inflammatory process. When inflammatory exudate was noted within the lumen of a sweat-gland coil, some break in the integrity of the connective-tissue sheath about the coil usually could be found (Fig. 4).

Treatment and Results

In order to evaluate the results of surgical treatment of hidradenitis suppurativa, follow up information was sought from the 64 patients who had been so treated. Information was obtained from 38 patients, the remaining 26 being lost to follow up.

The surgical treatment in most cases consisted of excision of the sites involved by hidradenitis. This was usually accomplished in one stage, but two-stage procedures were used in 15 cases and three-stage operations were necessary in six instances.

Eight of the 38 patients with follow up (21 per cent) had no subsequent symptoms; all these patients were followed for eight years or more. Twelve patients (32 per cent of those with follow up) had mild symptoms after operation but required no further treatment. The minimal period of follow up in this group was nine years except for one patient who was followed for only two months and another for three years. Seventeen patients (45 per cent of those followed) required further treatment after surgical intervention at the clinic. This subsequent therapy consisted of one or more further surgical procedures in almost all cases. The remaining patient died two years after excision of a grade 2 squamous cell epithelioma of the buttock; this patient had a 32-year history of hidradenitis.

Two deaths from cardiac disease were noted eight and nine years, respectively, after treatment at the clinic. Another patient died eight years after treatment at the clinic, the cause of death being reported as intraperitoneal infection secondary to perineal infection.

Summary and Conclusions

A study has been made at the Mayo Clinic on 117 patients who had hidradenitis suppurativa of the perianal region. Surgically removed tissue was available for study in 64 of these patients.

Perianal hidradenitis suppurativa occurs in males nearly twice as often as it does in females. The perianal region is involved in approximately one of every six cases of hidradenitis; the perianal region is involved alone in one of every 13 cases. Hidradenitis suppurativa may involve the neck, face, scalp, extremities, shoulders and abdomen, in addition to the usual sites.

Anal fistula secondary to hidradenitis is rare. Hidradenitis in the post sacral area is easily confused with pilonidal disease, especially if other parts of the body are not involved. Hidradenitis of the perianal region is usually not sharply confined to that region.

Malignant degeneration occasionally oc-
occurs as a complication of long-standing hidradenitis.

Hidradenitis tends to recur in spite of surgical treatment.

The apocrine sweat glands apparently are not of great importance in the pathogenesis of hidradenitis suppurativa; their involvement appears to be coincidental.

The cause and pathogenesis of hidradenitis suppurativa are not well understood. The disease apparently has some relationship to endocrine activity, and it appears likely that common etiologic factors are present in hidradenitis and acne. Other than the presence of apocrine glands, local conditions exist that might help to explain the typical distribution of hidradenitis suppurativa. Among these are the frequent apposition of cutaneous surfaces, excessive local moisture, heat and friction, and the difficulty of maintaining cleanliness in the regions frequently involved.

The treatment of hidradenitis of the perianal region is somewhat discouraging because many patients continue to experience recurrent activity of the disease even after careful excision of all active sites. To attempt to excise all of the skin in which recurrent activity might appear is out of the question, so one must be content to explain to the patient that future periodic surgical care is a possibility. Until the cause and pathogenesis of the disease are better understood, it is doubtful that the prognosis will improve.

References


PERIANAL HIDRADENITIS SUPPURATIVA


