Hidradenitis suppurativa: a disease with male predominance in Tunisia

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Background: hidradenitis suppurativa (HS) is a chronic inflammatory, suppurating, fistulizing, and scar-producing disease of apocrine gland–bearing skin. The diagnosis is primarily clinical, based on the presence of both sinus tracts and abscesses with a characteristic distribution.

Objective: Review of epidemiological, clinical, and prognostic characteristics of HS and discussion of the etiopathogenic aspects of this chronic problem.

Patients and methods: We retrospectively report all cases of HS followed at the Department of Dermatology between January 1985 and December 2008.

Results: Eleven patients (10 male and 1 female), with a mean age of 35.2 years (range 21–53 years) at HS diagnosis were followed for HS. The average age of disease onset was 23.9 years. The median delay between onset of symptoms and diagnosis was 144 months (range 1–408 months). Clinical features showed inflamed discharging papules or nodules, painful tender erythematous nodules, and double-ended comedones. The disease mainly affected the axillary, anal, perineal, and genital areas. Histologically, dermal features showed active folliculitis or abscess, sinus tract formation, fibrosis, and granuloma formation. Pathological associations (Darier's disease and Down syndrome) were noted in two patients. Treatment consisted of antibiotics in eight patients, retinoids (1 mg/kg/day) in three patients, and surgery in three patients. The mean follow-up was 13 months (range 2–30 months). Recurrence of lesions was observed in all patients approximately 1 month after treatment withdrawal. The Down syndrome patient developed vaginal hydrocele of the testis as a complication of his staphylococcic ulcers. In all cases healing occurred with substantial scarring.

Discussion: An obvious male predominance was noted in our patients as well as a delay in the diagnosis of HS, which could be explained by non-recognition of the disease by non-specialists that see the patients at the primary-care level.
Introduction

Hidradenitis suppurativa (HS), also known as Verneuil's disease or acne inversa, is a chronically relapsing inflammatory skin disease characterized by recurrent draining sinuses and abscesses, predominantly in skin folds with terminal hairs and apocrine glands. It was first described by Velpeau in 1839 then by Verneuil (2) in 1854, who associated it with the sweat glands. HS was then classified as a member of the follicular occlusion triad, along with acne conglobata and dissecting cellulitis of the scalp (3). In 1975, pilonidal sinus was added to this triad, forming the follicular occlusion tetrad (4). In 1989, Plewig and Steger (4) introduced the term acne inversa based on the follicular origin of the disease.

HS is usually diagnosed clinically. It has a chronic course and may be extremely painful and severely debilitating. This article reviews the epidemiological, clinical, and prognostic characteristics of HS.

Patients and methods

We retrospectively collected all cases of HS clinically diagnosed at the department of dermatology of La Rabta hospital, Tunis, between January 1985 and December 2008.

Relevant data included age, sex, delay between onset of symptoms and diagnosis, extent of disease, frequency of exacerbations, histological exam, and response to different treatment modalities.

Results

Eleven patients (10 males and 1 female) were referred for HS. The average patient’s age was 35.2 years (range 21–53 years) and the average age of disease onset was 23.9 years. At the time of the survey, patients had suffered average disease duration of 144 months (range 1–408 months). No family history of HS was noted in our patients. All patients had multifocal involvement. Clinical features showed inflamed discharging papules or nodules, painful tender erythematous nodules, and double-ended comedones (Figs. 1–3). The disease affected mainly the axillary, anal, perineal, and genital areas (Table 1). All patients were assessed according to Hurley’s clinical classification and eight of them were evaluated according to the Sartorius severity score (Table 2). In six patients, histological findings showed active folliculitis or abscess, sinus tract, fibrosis, and granuloma formation. Bacterial samples realized in seven cases revealed methi-S Staphylococcus aureus in all cases. Pathological
Table 1. Clinical and prognostic characteristics of HS patients.

<table>
<thead>
<tr>
<th>Patient data</th>
<th>Description</th>
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| Case 1       | Age: 41  
Sex: M  
Onset age: 25  
Duration: 16 yr  
Follow-up: 18 mo  
Personal history: Smoking, acne conglobata, Darier’s disease  
Aspect of lesions: Cysts, comedones, discharging sinuses  
Topography: Face, back, axillary and perineal areas  
Treatment: Acitretin 1 mg/kg/d  
Evolution: Improvement +++, relapses at retinoid withdrawal |
| Case 2       | Age: 29  
Sex: M  
Onset age: 21  
Duration: 8 yr  
Follow-up: 3 mo  
Personal history: Smoking  
Aspect of lesions: Nodules, boils, abscesses, ulcers, discharging sinuses, fibrosis, pustules, comedones  
Topography: Axillary, inguinal, perianal areas  
Treatment: Doxycyclines 200 mg/day, antiseptics, surgery  
Evolution: Improvement +, frequent relapses, lost to follow-up |
| Case 3       | Age: 28  
Sex: M  
Onset age: 20  
Duration: 8 yr  
Follow-up: 4 mo  
Personal history: Smoking  
Aspect of lesions: Abscesses, crypts, pustules, comedones  
Topography: Perianal, axillary areas  
Treatment: Oxacillin, antiseptics  
Evolution: Improvement +, lost to follow-up |
| Case 4       | Age: 21  
Sex: M  
Onset age: 18  
Duration: 3 yr  
Follow-up: 30 mo  
Personal history: Obesity, Down syndrome, hydrocele testis  
Aspect of lesions: Abscesses, sinus tract fistulae, nodules, comedones  
Topography: Axillary folds, groin, perianal, gluteal crease, perineum  
Treatment: Doxycyclines 200 mg/day, antiseptics  
Evolution: Improvement +++, relapses at antibiotic withdrawal |
| Case 5       | Age: 43  
Sex: M  
Onset age: 28  
Duration: 15 yr  
Follow-up: 6 mo  
Personal history: Diabetes type 2  
Aspect of lesions: Sinus tract fistulae, ulcerations  
Topography: Inguino-scrotal, perineum, gluteal areas  
Treatment: Doxycyclines 200 mg/day, antiseptics  
Evolution: Improvement ++, relapses at antibiotic withdrawal |
| Case 6       | Age: 53  
Sex: F  
Onset age: 19  
Duration: 34 yr  
Follow-up: 24 mo  
Personal history: Acne vulgaris  
Aspect of lesions: Fistulizing lesions, folliculitis  
Topography: Axillary, inguinal, perianal areas  
Treatment: Isotretinoin 1 mg/kg/day, topical erythromycin  
Evolution: Improvement +++, relapses at retinoid withdrawal |
| Case 7       | Age: 27  
Sex: M  
Onset age: 45  
Duration: 1 mo  
Follow-up: 3 mo  
Personal history: –  
Aspect of lesions: Suppurating lesions  
Topography: Axillary folds  
Treatment: Oxacillin antiseptics  
Evolution: Improvement +, lost to follow-up |
| Case 8       | Age: 45  
Sex: M  
Onset age: 45  
Duration: 2 mo  
Follow-up: 6 mo  
Personal history: Smoking  
Aspect of lesions: Bridle scars, folliculitis, sebaceous cysts  
Topography: Axillary folds, perianal area  
Treatment: Doxycyclines 200 mg/day, antiseptics, topical erythromycin  
Evolution: Improvement +++, relapses at antibiotic withdrawal |
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**Case 9**

Age: 38
Sex: M
Onset age: 24
Duration: 14 yr
Follow-up: 20 mo

Personal history: Smoking
Aspect of lesions: Bridle scars, purulent fistulas, abscess, keloidal atrophic scars
Topography: Axillary folds, perianal, intergluteal regions
Treatment: Rifampicin, antiseptics, topical erythromycin
Evolution: Improvement ++, relapses at antibiotic withdrawal

**Case 10**

Age:
Sex:
Onset age:
Duration: yr
Follow-up: 6 mo

Personal history: Acne vulgaris
Aspect of lesions: Nodulo-cystic lesions, comedones, papulo-pustules, bridles, interconnecting abscesses, keloidal atrophic scars
Topography: Face, back, axillary folds
Treatment: Doxycyclines 200 mg/day, antiseptics, surgery
Evolution: Improvement ++, relapses at antibiotic withdrawal

**Case 11**

Age:
Sex:
Onset age:
Duration: yr
Follow-up: 6 mo

Personal history: Acne vulgaris, smoking
Aspect of lesions: Nodulo-cystic lesions, comedones, papulo-pustules, interconnecting abscesses
Topography: Axillary folds, perianal, intergluteal regions, face
Treatment: Isotretinoin 1 mg/kg/day, surgery
Evolution: Improvement ++, relapses at retinoid withdrawal

**Table 2. Clinical classification of HS in our patients**

<table>
<thead>
<tr>
<th>Case</th>
<th>Hurley’s clinical classification (stages)</th>
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<tr>
<td>1</td>
<td>I</td>
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<td>10</td>
<td>III</td>
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<tr>
<td>11</td>
<td>III</td>
<td>42</td>
</tr>
</tbody>
</table>

associations (Darier’s disease and Down syndrome) were noted in two patients.

Treatment consisted of antibiotics (oxacillin, doxycyclin, or rifampicin) in eight patients, retinoids (1 mg/kg/day) in three patients, and local surgery of discharging nodules and abscesses in three patients. Mean follow-up was 16 months (range 2–36 months). Follow-up was marked with recurrence of lesions in all patients approximately 1 month after treatment withdrawal, leading to cyclic readministration of antibiotics or retinoids. The Down syndrome patient developed vaginal hydrocele of the testis as a complication of staphylococcic ulcers. Healing occurred with substantial scarring (keloidal atrophic scars) in three patients. One patient had limited surgical excision of keloidal scars.
Discussion

Hidradenitis suppurativa (HS) is a rare chronic suppurative condition that is associated with significant morbidity.

The exact incidence of HS is uncertain, but the literature suggests from 1 in 300 to 600 (4–6). HS is probably more common than once thought, but the diagnosis is frequently ignored or missed.

The disease seems to be more common in females, with reported female:male ratios ranging from 2:1 to 5:1 (4–6). In our series, a marked male preponderance with reported female:male ratios ranging from 2:1 to 5:1 (4–6). In our series, a marked male preponderance is noted with a sex ratio (M/F) of 10/1.

The average age of onset of HS, as in our patients, is 23 years (4–6). In less than 2% of cases, the disease appears before age 11 and, in extremely rare cases, it occurs after menopause (7). In addition, the disease tends to ease or subside in women after menopause, probably in relation to hormonal changes (7, 8).

An increased frequency of HS is observed in blacks, possibly because blacks have a greater density of apocrine glands than whites (4, 8). In our series, all patients had dark phenotype (IV or V). HS has polymorphic clinical and evolutive courses. Disease onset is insidious, and early symptoms may include discomfort, itching, erythema, burning, and hyperhidrosis. Occlusion of a hair follicle results in large multilobulated comedones, then nodules or cysts (3, 9, 10). The cyst may rupture spontaneously, leading to purulent discharge and chronic draining sinuses. Otherwise indurated inflammatory deep abscesses may occur. The lesions then heal with fibrosis, leading to hypertrophic or keloid scarred skin and subcutaneous tissues (6, 9, 10).

The clinical course varies from occasional axillary lesions to diffuse abscess formation in multiple sites. The sites of predilection of lesions are genitofemoral areas in women and perianal involvement in men. No gender predilection is seen in the axillary lesions (4, 6).

Patients are evaluated using the simplified Hurley’s clinical classification (9) or the Sartorius severity score (10).

In the case of suspicious lesions as well as perianal involvement, biopsies should be performed to exclude the possibility of coexisting cancer, and Crohn’s disease should also be considered. In the majority of specimens, the histological examination reveals follicular involvement, including poral occlusion and folliculitis. Apocrine glands are the dominant histological feature found in only a small number of specimens (11). Furthermore, a paucity of apocrine glands is noted in the genitofemoral region. This finding supports the theory that apocrine gland inflammation is not the pathogenetic mechanism of HS, but rather a secondary manifestation of follicular involvement (6, 12).

Bacteriological analyses of HS have also been carried out. Deep needle aspiration and the carbon dioxide (CO2) laser method have revealed Staphylococcus aureus and coagulase-negative staphylococci to be the most commonly found bacteria (13, 14). Imaging studies including MRI and ultrasound have otherwise been used to determine locoregional involvement (15, 16). In our patients, bacteriological analysis revealed Staphylococcus aureus and coagulase-negative staphylococci in seven patients tested.

The etiology of HS is still being debated. Although several studies have failed to demonstrate human lymphocyte antigen (HLA) associations (17), others have suggested an autosomal dominant mode of inheritance (18). Otherwise, hyperandrogenism, obesity via occlusion and maceration, heat, humidity and friction from clothing, smoking, lithium, chemical irritants, and oral contraceptives may be associated with HS, possibly as triggering factors (19–24). The bacterial infection is thought to occur secondary to the disease process (25). In addition, HS has been reported to coexist with other skin diseases that show poral occlusion; for example, Fox-Fordyce disease, pityriasis rubra pilaris, steatocystoma multiplex, and Dowling-Degos disease (4). Associations with Down syndrome, Behçet’s disease, acanthosis nigricans, pyoderma gangrenosum, pyoderma vegetans, and arthropathy have also been described (4). Regarding the relationship with Crohn’s disease (CD), some have hypothesized that perianal involvement of HS appears to be another cutaneous manifestation of CD (26). Although foreign body--type granulomas are a common finding in HS, the presence of discrete epithelioid granulomas in the dermis away from the site of active inflammation is unusual and should alert the pathologist to the possibility of a systemic granulomatous disease such as CD (26, 27). In our study, pathological associations have included Down syndrome and Darier’s disease, and to our knowledge there has been no previous report of such an association in the literature. The association between diabetes and HS as in patient 5 is sporadically described (28). HS is often a diagnostic challenge, and in the early stages the differential diagnosis includes a painful nodule, abscess, furuncle, carbuncle, lymphadenitis, and ruptured inclusion cyst (4, 27). In later stages, when inflammation of more than one gland is present, the differential includes lymphogranuloma venereum, donovanosis, scrofuloderma, tuberculous gumma, actinomycosis, sinus tracts, and fistulas occurring with ulcerative colitis and regional enteritis (4, 27, 28).
HS is a recurrent disease with a chronic and progressive clinical course. When measured by the Dermatology Life Quality Index (DLQI), patients experience a significant degree of morbidity, with the highest scores resulting from pain caused by disease. Additionally, quality of life seems to be lower than in other dermatologic diseases such as urticaria, psoriasis, atopic dermatitis, and neurofibromatosis (29).

Potential complications include dermal contraction, local or disseminated infection, lymphedema caused by lymphatic injury from inflammation and scarring, rectal or urethral fistulas, restricted limb mobility from scarring, and arthritis secondary to inflammatory injury (4). Reports of squamous cell carcinoma following chronic lesions of HS have been described (30). Other rare but serious complications of HS are bacterial meningitis, bronchitis, pneumonia, and systemic amyloidosis (4). In our study, the Down syndrome patient developed vaginal hydrocele of the testis as a complication of staphylococcal ulcers. Two patients presented severe keloidal and atrophic scars.

There is no single effective treatment for HS. In mild cases we can begin with conservative measures such as warm baths, hydrotherapy, cryotherapy, and topical cleansing agents to reduce bacterial load (31). Nonsteroidal anti-inflammatory drugs may alleviate pain as well as inflammation. Antibiotics, although not proven to be effective, are the mainstay of medical treatment, especially for lesions suspected of being superinfected. The only topical antibiotic that has been proven effective in a randomized controlled trial is clindamycin (32). Antistaphylococcal agents are best for axillary disease, and more broad-spectrum coverage is better for perineal disease. Dicloxacillin, erythromycin, tetracycline, and minocycline have been used with various results (32). Other hormonal medications have been tried (oral contraceptive agents that contain a high estrogen-to-progesterone ratio, finasteride in males) with various responses (33, 34). Oral retinoids (isoretinoin, acitretin, etretinate) have also been used with prolonged remissions (35, 36). Systemic or intralesional corticosteroids, immunosuppressant agents, and intramuscular human immunoglobulin are other treatment possibilities (6, 31). Significant clinical improvement with long remissions was also reported after administration of infliximab and etanercept (37, 38). Radiotherapy has been investigated as a potential treatment option (39). Aminolevulinic acid photodynamic therapy for HS has also been reported as an interesting alternative (40). Otherwise, carbon dioxide laser has been used in conjunction with second-intention healing to provide relief for a few patients (41).

Finally, although incision, drainage, and exteriorization of individual lesions may be useful in some instances, radical surgical excision at the earliest recognized stage remains a mainstay of therapy. Postoperative recurrence is common after incision and drainage with limited surgical excision (42).

In our study, nine patients had antibiotics (1- to 3-month cures) and three patients had systemic retinoids (1 mg/kg/day) with relapses after the first month of treatment withdrawal. Three patients had incision, drainage, and exteriorization of individual lesions, and one had limited surgical excision of keloidal scars.

**Conclusion**

HS remains a challenging disease for patients and physicians. In addition to treating the physical illness, it is crucial to acknowledge and treat the psychological burden associated with the disease. Because of the areas of the body that are affected, the malodorous discharge, the chronic discomfort, and the general unsightliness of the disease as well as years of inadequate treatment, HS may lead to frustration, depression, and isolation.

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