

Hidradenitis suppurativa: a debilitating disease with male predominance in Tunisia

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Citation

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Abstract

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: to review epidemio-clinical and prognostic characteristics of HS and discuss etiopathogenic aspects of this chronic affection. Patients and methods: we retrospectively report all cases of HS followed at the Department of Dermatology between January 1985 and December 2008. Results: 10 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 (1 month - 408 months). Clinical features showed inflamed discharging papules or nodules, painful tender erythematous nodules and double ended comedons. The disease affected mainly 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's syndrome) were noted in 2 patients. Treatment consisted of antibiotics in 8 patients, retinoids (1 mg/kg/day) in 3 patients and surgery in 3 patients. Mean follow-up was 13 months (range 2-30 months). Recurrence of lesions was observed in all patients approximately 1 month after treatment withdrawal. Down's syndrome patient developed a vaginal hydrocele testis as a complication of his staphylococcal 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 the non specialists who see the patients in the front line.

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 that carry terminal hairs and apocrine glands. It was first described by Velpeau in 1839 then by Verneuil (1) in 1854 that 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 (2). In 1975, pilonidal sinus was added to this triad, forming the follicular occlusion tetrad (3). In 1989, Plewig and Steger (3) 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.

Objective: to review epidemio-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 survey, patients had suffered average disease duration of 144 months (range 1 month - 408 months). All patients had multifocal involvement. No family history of HS was noted in our patients. Clinical features showed inflamed discharging papules or nodules, painful tender erythematous nodules and double ended comedons (figures 1,2,3).

Figure 1

Figure 1: discharging nodules and folliculitis of the axillary's fold



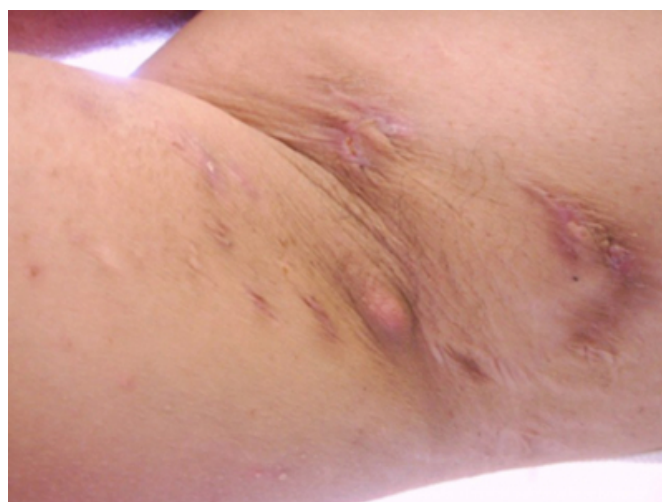
Figure 2

Figure 2: purulent draining sinuses and discharging nodules



Figure 3

Figure 3: bridle hypertrophic scars



The disease affected mainly the axillary, anal, perineal and genital areas (table 1).

Figure 4

Table 1: clinical and prognostic characteristics of our patients

case	AGE/SEX	AGE AT ONSET	HS DURATION (years)	PERSONAL HISTORY	ASPECT OF LESIONS	TOPOGRAPHY	TREATMENT	EVOLUTION/ follow up duration
1	41/M	25	16	-Smoking -acne conglobata -Darier's disease	Cysts, comedons, discharging sinuses	Face, back, axillary and perineal areas	Acitretin 1mg/kg/d	Improvement +++ Relapses at retinoid withdrawal/ 18 months
2	29/M	21	8	smoking	Nodules, boils, abscesses, ulcers, discharging sinuses, fibrosis, pustules, comedons	Axillary, inguinal, perineal areas	Doxycyclines 200mg/day antiseptics surgery	Improvement + Frequent relapses Lost to follow-up / 3 months
3	28/M	20	8	smoking	Abscesses, crypts, pustules, comedons	Perianal, axillary areas	Oxacillin antiseptics	Improvement + Lost to follow-up/ 4 months
4	21/M	18	3	-Obesity -Down's syndrome -hydrocele testis	Abscesses, sinus tract fistulae, nodules, comedons	Axillary folds, groin, perianal, gluteal crease, perineum	Doxycyclines 200mg/day antiseptics	Improvement +++ Relapses at antibiotic withdrawal/ 30 months
5	43/M	28	15	Diabetes type 2	Sinus tract fistulae, ulcerations	Inguino-scrotal, perineum, gluteal areas	Doxycyclines 200mg/day antiseptics	Improvement ++ Relapses at antibiotic withdrawal/ 6 months
6	53/F	19	34	Acne vulgaris	Fistulising lesions, folliculitis	Axillary, inguinal, perianal areas	Isotretinoin 1mg/kg/day topical erythromycin	Improvement ++ Relapses at retinoid withdrawal/ 24 months
7	27/M	27	1 month	-	Suppurating lesions	Axillary folds	Oxacillin antiseptics	Improvement + Lost to follow-up/ 3 months
8	45/M	45	2 months	smoking	-bridle scars -folliculitis -sebaceous cysts	-Axillary folds -Perianal area	Doxycyclines 200mg/day antiseptics topical erythromycin	Improvement ++ Relapses at antibiotic withdrawal/ 6 months
9	38/M	24	14 years	smoking	-bridle scars -purulent fistulas -abscess -Keloidal atrophic scars	-Axillary folds -Perianal, intergluteal regions	Rifampicin antiseptics topical erythromycin	Improvement ++ Relapses at antibiotic withdrawal/ 20 months
10	28/M	12	16 years	Acne vulgaris	-nodulo-cystic lesions -comedones -papulo-pustules -bridles -interconnecting abscesses -Keloidal atrophic scars	-Face -Back -Axillary folds	Doxycyclines 200mg/day antiseptics surgery	Improvement ++ Relapses at antibiotic withdrawal/ 6 months
11	38/M	23	15 years	Acne vulgaris smoking	-nodulo-cystic lesions -comedones -papulo-pustules -interconnecting abscesses	-Axillary folds -Perianal, intergluteal regions - Face	Isotretinoin 1mg/kg/day surgery	Improvement ++ Relapses at retinoid withdrawal/ 6 months

All patients were assessed according to Hurley's clinical classification and 8 of them were evaluated according to the Sartorius severity score (Table 2).

Figure 5

Table 2: clinical classification of HS in our patients

case	Hurley's clinical classification (stages)	Sartorius score
1	I	-
2	III	-
3	II	35
4	III	90
5	III	56
6	II	38
7	II	19
8	III	85
9	II	33
10	III	-
11	III	42

In 6 patients, histological findings showed active folliculitis or abscess, sinus tract, fibrosis, and granuloma formation. Bacterial samples realized in 7 cases revealed a methi S staphylococcus aureus in all cases. Pathological associations (Darier's disease and Down's syndrome) were noted in 2 patients.

Treatment consisted of antibiotics (oxacillin, doxycyclin or rifampicin) in 8 patients, retinoids: (1 mg/kg/day) in 3 patients and local surgery of discharging nodules and abscesses in 3 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. Down's syndrome patient developed a vaginal hydrocele testis as a complication of staphylococcic ulcers. Healing occurred with substantial scarring (Keloidal atrophic scars) in 3 patients. One patient had a limited surgical excision of keloidal scars.

DISCUSSION

Hidradenitis suppurativa (HS) is a rare chronic suppurative condition which is associated to significant morbidity.

The exact incidence of HS is uncertain, but the literature suggests from one in 300 to one in 600 (3-5). 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 to 5:1 (3-5). 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, like in our patients, is 23 years (3-5). In less than 2% of cases, the disease appears before age 11 years and in extremely rare cases, it occurs after menopause (6). Besides, the disease tends to ease or subside in women after the menopause, probably in relationship with hormonal changes (6,7).

An increased frequency of HS is observed in blacks, possibly because blacks have a greater density of apocrine glands than whites (3,7). In our series, all patients had dark phototype (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 multiheaded comedones, than in nodules or cysts (2,8,9). 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 (5,8,9).

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 (3,5).

Patients are evaluated using a simplified clinical classification "Hurley's clinical classification" (8) or the Sartorius severity score (9).

In case of suspicious lesions as well as perianal involvement, biopsies should be performed to exclude the possibility of coexisting cancer and crohn's disease (CD) should also be considered. In the majority of specimens, the histological examination reveals follicular involvement, including poral occlusion and folliculitis. Apocrinitis as the dominant histological feature is found in only a small number of specimens (10). 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 (5,11). Regarding the relationship with CD; 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 should alert the pathologist to the possibility of a CD (12).

Bacteriological analyses have also been realized in HS. Deep needle aspiration and 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 ultrasonography have been otherwise used to determine locoregional involvements (15). In our patients, bacteriological analysis revealed Staphylococcus aureus and

coagulase-negative staphylococci in 7 tested patients.

The aetiology of HS is still under debate. While several studies have failed to demonstrate human lymphocyte antigens (HLA) associations (16), others have suggested an autosomal dominant mode of inheritance (17). Otherwise, a 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 (18-23). The bacterial infection is thought to occur secondary to the disease process (24). Besides, HS has been reported to coexist with other skin diseases that show poral occlusion, e.g., Fox–Fordyce disease, pityriasis rubra pilaris, steatocystoma multiplex and Dowling–Degos disease (3,5). Associations with Down’s syndrome, Behcet’s disease, acanthosis nigricans, pyoderma gangrenosum, pyoderma vegetans and arthropathy have also been described (3). In our study, pathological associations have included Down’s 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 (25). 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 (3,24). 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 (3,24,25).

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 highest scores obtained from pain caused by disease. Additionally, quality of life seems to be lower than other dermatologic diseases such as urticaria, psoriasis, atopic dermatitis and neurofibromatosis (26).

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 (3). Reports of squamous cell carcinoma following chronic lesions of HS have been described (27). Other rare but serious complications of HS are bacterial meningitis, bronchitis, pneumonia and systemic amyloidosis (5,28). In our study, Down’s syndrome patient has developed a vaginal hydrocele testis as a complication of

staphylococcal ulcers. Two patients have 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 (5).

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 (5,29). Carbon dioxide laser surgery constitutes also an interesting alternative of early HS lesions (30).

Antibiotics, although not proven to be effective, are frequently used, especially for lesions suspected of being infected (31). Hormonal medications (anti-androgen cyproterone acetate in conjunction with ethinylestradiol in females and the 5- α -reductase inhibitor, Finasteride in males) have been tried with variable results (32). Oral retinoids (isotretinoin, acitretin, etretinate) have showed prolonged remissions (5,33). Corticosteroids, immunosuppressants and human immunoglobulin are other treatment possibilities (5). Radiotherapy and Aminolevulinic acid photodynamic therapy have been investigated as a potential treatment option and recently, significant clinical improvements with long remissions were reported with infliximab and etanercept (5,34).

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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