# Chronic hidradenitis suppurativa in the inguinal, perineal and scrotal regions.
## A case report and review of the literature

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**Chronic hidradenitis suppurativa in the inguinal, perineal and scrotal regions. A case report and review of the literature**

*Hidradenitis suppurativa is a chronic, recurrent, debilitating disease that presents with inflamed lesions in the apocrine glands of the body. The most common locations are the axillary, inguinal and anogenital areas. Hidradenitis suppurativa is caused primarily by follicular occlusion with secondary involvement of the apocrine glands.*
*The authors report a case of 47-old-man with an 18-year history of multiple sclerosis complicated by spastic paraparesis, who presented with hidradenitis suppurativa in the inguinal, perineal, and scrotal areas which was treated by wide surgical excision. A review of the most recent literature is included.*

KEY WORDS: Abscess, Hidradenitis suppurativa, Verneuil's disease.

## Introduction

Hidradenitis suppurativa (HS) is a condition consisting of chronic and recurrent inflammation of the hair follicles located in the body folds, (in the axillary, inguinal, mammary, submammary, perianal and scrotal regions). It is characterized by subcutaneous nodules which evolve into fistulous tracks and recurrent abscesses.
Hidradenitis suppurativa is most often found in the axillae. The disease has a more insidious progression in the perianal and inguinoscrotal regions, where it occurs in 25% and 40% of patients respectively.
The hypothesis that the disease originates primarily in the apocrine glands appears to have been abandoned, although these glands may be one of the factors that contribute to the development of HS [1]. Changes in the immune system and a familiar component may be the primary factors implicated in the pathogenesis of HS, while secondary factors involved appear to be hyperandrogenism, bacterial colonization, poor hygiene, smoking, and obesity [2,3].
The disease was first described by the french physician Volpeau in 1839, but it was Verneuil, in 1854, who proposed the first hypothesis regarding its pathogenesis; purulent inflammation of the apocrine glands. This theory was accepted for over a century, and to such an extent that many authors used the term "disease of Verneuil" for the complex of symptoms associated with HS [4].
The association of the primary suppurative process of the apocrine sudoriferous glands and Verneuil's disease is based on the fact that the location of the lesions corresponds to that of these sweat glands. Some authors claim that these glands alone are responsible for the pathogenesis of HS, but others maintain that the pilosebaceous follicles are involved as well.

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The exact frequency of HS has not been determined, but in various case series was found to be 2-3 cases/1000, with the incidence increasing in the second to third decades of life, and prevalence higher among women (3:1), although the disease is rare in postmenopausal women [5,6].
The author's aim was to discuss correct management and treatment of patients with complicated HS based on a case report and a review of the most recent literature.

## Case report

G. P., a 47-year-old male smoker, suffering by multiple sclerosis diagnosed 18 years previously, later complicated by spastic paraparesis, under medical treatment. He complained of the intermittent appearance, since 3 years ,of numerous recurrent small, reddish nodules, both painful and tender, on the perineum and scrotum as well as in the gluteal regions. His condition had been complicated by fistulas and chronic, recurrent abscesses treated conservatively in other centers. Physical examination of the perineum, scrotum, and the gluteal regions revealed a vast erythematous, edematous, area of suppuration with nodular formations (abscesses) and many fistulous openings. Squeezing of the nodular formations elicited pain and purulent material was expressed. This clinical picture was compatible with a diagnosis of Hurley stage III HS with a Sartorius score of 106. After routine blood tests, a chest x-ray, an electrocardiogram and a complete cardiac examination, the patient underwent surgery. A total of 2 operations, 2 months apart, were required to treat all the abscesses and fistulas.
In the first operation the gluteoperineal region was treated. When the patient was in the lithotomy position over 20 external fistulous openings were found in the perineum and scrotum. After identification of the external orifice (posterolateral, seven o'clock) additional purulent material flowed out and the fistula track was followed and was found to be transphincteric with its internal orifice at six o'clock. Complete fistulectomy was performed to treat this fistula. There were numerous secondary tracks branching off it, some of which were laid open and some cauterized. The same treatment was used for a left posterolateral fistula. Five other fistula tracks were laid open. Hemostasis was achieved and the surgical wounds were medicated with iodoform gauze dressings (Fig. 1).
The surgical specimens examined had marked dermal and subcutaneous fibrosis with diffuse foci of infiltrating lymphocytes and plasma cells, especially around blood vessels, as in chronic HS.
In the second operation perineal and scrotal region was treated with removal of all fistula tracks and laid open of more complex. Hemostasis was achieved and the surgical wounds were medicated with iodoform gauze dressings (Fig. 2).
The patient's postoperative course after each operation was uneventful. Antibiotic and analgesic therapy was administered for 36-48 hours postoperatively.
The patient was discharged home on postoperative day 3 the first time and on postoperative day 1 after the

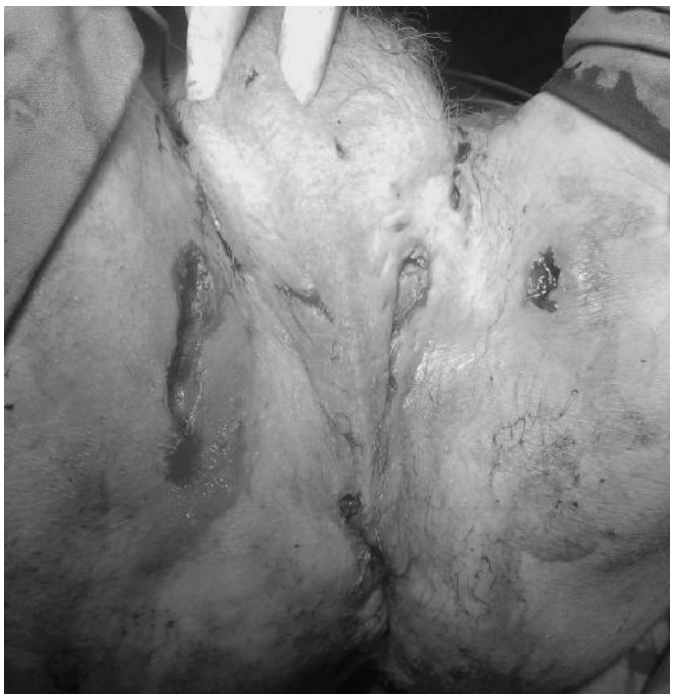

Fig. 1: Treatment of the gluteoperineal region. Healing will take place by secondary intention.

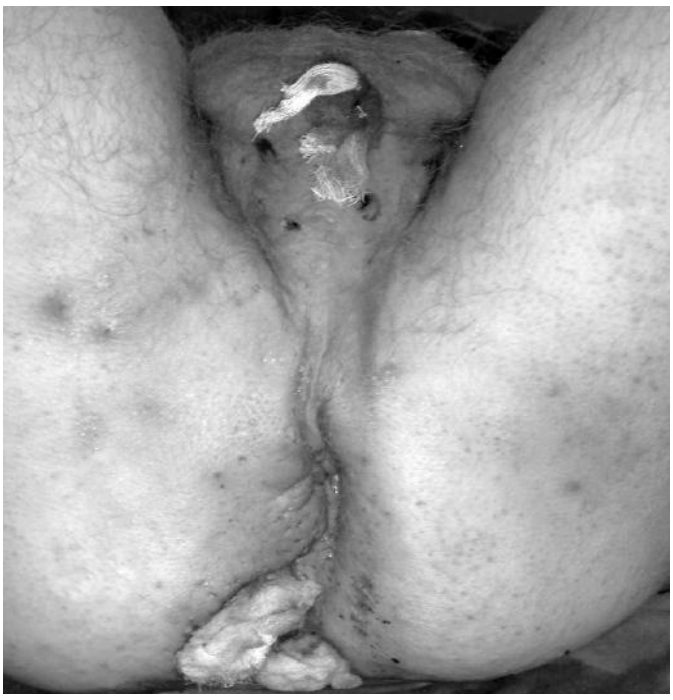

Fig. 2: Perineal and scrotal region treated.

second operation. He was initially followed up daily, then weekly and currently has monthly check-ups.

## Discussion

Hidradenitis suppurativa is a complex disease of uncertain etiology. It is characterized by a chronic, suppurative lesion with a tendency to fistulization and sclerosis, which develops from the subcutaneous tissue in regions in which apocrine glands are located. Diagnosis is based on the clinical evolution of the disease, since no characteristic features of HS can be identified on pathologic examination. In fact, the cutaneous and subcutaneous tissue comes to resemble an aspecific granuloma with abundant sclerosis, skin breakdown and the disappearance of hair follicles as well as sebaceous and sudoriferous glands.

Hidradenitis suppurativa does not pose any particular problems of differential diagnosis. Pathologies to be ruled out are; tuberculosis, perianal amebiasis, a perianal eosinophilic granuloma, actinomycosis, a suppurative pilonidal cyst, conglobated acne, Crohn's disease, and some tumors of the sudoriferous glands. The latter represent a special challenge for various reasons, some intrinsic, such as their relative rarity, and some extrinsic, relating to problems of nomenclature and classification. Since the sudoriferous glands are commonly classified as eccrine or apocrine, many authors have suggested classifying the tumors on the basis of these properties, but this idea was soon abandoned due to the confusion that occurred when both types (eccrine and apocrine) were found in the same tumor. A simpler classification, into rare and not so rare benign tumors (adenomas) and malignant ones (carcinomas) is preferred (Table I).

For these reasons treatment must be adjusted to the patient's clinical condition which is evaluated by means of certain parameters. The scores most widely used to evaluate HS patients are the Hurley classification (Table II), and the Sartorius score modified by Revuz (Table III) [7-10]. The former is easier and quicker to use, while the latter is a more complete but more laborious assessment of the patient's condition. Even though neither has obtained unanimous recognition or validation, they can be used as an initial indication for surgical or conservative treatment.

Klipfel et al maintain that immediate smoking cessation and weight loss are essential for HS patients. Although from a general standpoint this is beneficial because of the association of smoking and excess body weight with disease, neither seems to be a risk factor for HS [11]. Aspects of personal hygiene that could be beneficial are reduction of inflammation and local skin irritation and the use of antibacterial soap. If this does not lead to improvement in the 3-6 months that follow, medical therapy seems indicated for the initial stages of the disease.

The antibiotics clindamycin (300mg twice daily) and rifampicin (600mg once daily), singly or in combination, have remained first-line treatment for the first two stages of HS [11-12].

Hormonal therapy, together with or following antibiotic therapy, has also been proposed. The antiandrogens (ciproterone acetate and etinilestradiol), and finasteride seem to be the drugs that produce the best results [14,15].

The retinoids are another class of drugs used to treat HS. Isotretinoin, used successfully against acne, does not appear to provide the same results in patients with Verneuil's disease. However, in recent studies, another drug, Acetretin (25 mg twice daily), has been shown to be more effective.

The use of steroids, both topical and systemic, is con-

TABLE I - *Classification of the sudoriferous glands tumors.*

| **Adenomas** | **Carcinomas** |
|---|---|
| Cystic/microcystic structure | Porocarcinoma |
| – Hidrocystoma | Hidradenocarcinoma |
| – Syringoma | Ductal carcinoma |
| – Tubular (or papillary) adenoma | Apocrine carcinoma |
| | Syringomatous carcinoma (microcystic carcinoma) |
| Papillary structure | |
| – Hidradenoma papilliferum | Rare forms: |
| – Syringocystadenoma papilliferum | |
| Structure: solid/cystic | Syringocarcinoma papilliferum |
| – Chondroid syringoma | Mixed malignant tumor |
| – Nodular/cystic hidradenoma | Spiradenocarcinoma |
| – Clear cell poroma | Cylindrocarcinoma |
| – Spiradenoma | Adenoidocystic carcinoma |
| – Cylindroma | Mucinous carcinoma |
| – Eccrine poroma | Extramammary Paget's disease |

TABLE II - *Hurley Staging System.*

| Stage | Characteristics |
|---|---|
| I | Abscess formation, single or multiple, without sinus tracts or cicatrization. |
| II | Recurrent abscesses, with sinus tract formation and cicatrization; single or multiple, widely separated lesions. |
| III | Diffuse or near-diffuse involvement, or multiple interconnected tracts and abscesses across entire are. |

troversial. Although on the one hand they reduce local inflammation, on the other hand they favor the proliferation of bacteria. The same is true of all other immunosuppressant drugs, some of which (cyclosporin) are frequently used in medical therapy of HS [13].

The role of radiotherapy, must also be clearly defined, since in recent studies the results of radiotherapy have been widely divergent. Its use must be limited to selected cases, and physicians must keep in mind that administering radiation as a treatment for non-neoplastic disease exposes the patient to a heightened risk of mutations [16,17].

If HS does not respond to medical treatment and improved hygiene, and becomes chronic, with severe manifestations, surgery, though not obligatory, is a wise choice, given the natural course of the disease and its clinical evolution [18,19].

There is still controversy regarding the choice of surgical treatment. Various factors, such as disease extent, the region(s) of the body involved, and whether the lesions are acute or chronic, can affect the outcome.

Small nodules and abscesses can be treated with simple drainage, but if this is not followed by extended excision of the fistula tracks, recurrence is almost certain to occur after an interval of 3 months on average.

Some authors maintain that if skin involvement is limited, local excision of the fistula tracks with marsupialization may be indicated. However, recent studies show that the recurrence rate after this treatment option is around 50% [20-24].

Therefore, the best surgical approach, even for clinically limited lesions, seems to be a wide excision of the lesion(s), including approximately 0.5-2 cm of healthy tissue at the lateral and deep margins. The block of tissue removed must include both the diseased area and the adjacent zone containing apocrine glands. This significantly reduces the risk of local recurrence [20,25,26].

We believe, as do many other authors, that healing by secondary intention, with the application of iodoform gauze dressings and periodic medications, is more than adequate treatment, both because there are often very extended lesions, and, more importantly, because of the contamination and inflammation present [22,27].

Controversy exists regarding methods of surgical wound treatment and the possibility of direct closure using covering flaps or grafts. Although this technique can be applied to lesions in areas of the body like the axillae, it is unsuitable and contraindicated for other areas like the inguinoperineal region [28-31].

Good results have been obtained in the treatment of moderate and severe forms of HS with carbon dioxide lasers and healing by secondary intention, a method which appears to reduce scarring and postoperative pain [26,32-34].

Potential complications of surgery are tissue fibrosis, excessive and retractive scarring, especially in the axillae. However, if there is anogenital disease, the most greatly feared complications are anal, rectal, or urethral fistulas.

In the case reported here, the patient had very advanced HS which was complicated by comorbidity (multiple sclerosis with spastic paraplegia) that had kept him bedridden thus aggravating his general and local condition.

The fistulae were removed in toto, with a good-sized margin of healthy tissue, sent to the pathologist for the

TABLE III - *Sartorius score modified by Revuz.*

| Characteristics | No. | Coefficient | Total |
|---|---|---|---|
| 1. Anatomical region involved: armpit, breast, inguinofemoral, perianal and perineal | | X 3 | |
| 2. Lesions: - Nodules | | X 2 | |
| - Abscess or fistulas | | X 4 | |
| - Hypertrophic scars | | X 1 | |
| - Other (eg, folliculitis, pustules) | | X 0.5 | |
| 3. Longest distance between two relevant lesions or size if only one lesion < 5 cm = 2; < 10 cm = 4; > 10 cm = 6; no active lesion = 0) | | X 1 | |
| 4. Are all lesion clearly separated by normal- appearing skin? (yes = 0; no = 6) | | X 1 | |
| **Total** | | | |

definitive histology examination, (although cases of chronic HS associated with squamous cell carcinoma are very rare in the literature), The more complex fistulas were laid open, the affected tissue was excised, and the wound was left to heal by secondary intention [35,36].

## Conclusions

Hidradenitis suppurativa is a little known disease and for this reason there is no unified approach to treatment. Helping patients modify their personal hygiene can, without doubt, be beneficial, especially in the initial phases of the disease. The medical treatment indicated for mild and acute forms of HS is a useful adjunct to surgical treatment, with wide excision of the lesions and healing by secondary intention, which seems the best treatment choice if the disease has become chronic/recurrent, or unresponsive to medical therapy, or is a moderate/severe form of HS.
We believe that simple drainage or local excision are treatments that should be reserved for patients with limited lesions and no concomitant pathologies that aggravate the condition.


## Riassunto

L'idrosadenite suppurativa (IS) è una patologia infiammatoria cronica e ricorrente dei follicoli piliferi caratterizzata, a livello delle pieghe del corpo (ascelle, inguine, regione mammaria e sotto-mammaria, regione perianale e scrotale) da noduli sottocutanei che evolvono in tramiti fistolosi ed ascessi ricorrenti.
Gli autori riportano il caso clinico di un uomo di 47 anni, affetto da sclerosi multipla complicata da paraparesi spastica ed idrosadenite suppurativa a livello delle regioni perineali, inguinali e scrotali, trattato chirurgicamente con buoni risultati.
Dopo una revisione della letteratura, gli autori analizzano, non tralasciando l'aspetto igienico-comportamentale, le varie tipologie di trattamento medico e chirurgico, focalizzando l'attenzione sull'escissione ampia delle lesioni per ridurre al minimo il rischio di recidiva.


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