

Hidradenitis Suppurativa: Case Report

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DOI: <https://doi.org/10.36347/sjmcr.2025.v13i03.006>

| Received: 06.12.2024 | Accepted: 15.01.2025 | Published: 06.03.2025

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Abstract

Case Report

Verneuil's disease or hidradenitis suppurativa is a chronic inflammatory and suppurative, fistulizing and sclerosing disease of the pilosebaceous follicles of the anatomical regions rich in apocrine glands. The disease was named by Verneuil, a French surgeon, who in 1864 argued that the initial phenomenon was an inflammation of the sweat glands. This is a rare condition whose incidence has not been clearly established; the prevalence varies from 0.03% to 8% in industrialized countries. The pathophysiology of Verneuil's disease is still subject to much controversy. Studies demonstrate the presence of multiple risk factors associated with Verneuil's disease: smoking and obesity are the two main risk factors. Its positive diagnosis remains clinical, however there are many differential diagnoses, hence the interest in the para-clinical assessment and in particular the anatomo-pathological study to allow the discernment of these diagnoses. Although the prognosis is rarely life-threatening, some complications can be serious, both locally (malignant degeneration) and generally, where we note the importance of the psychological and socio-professional impact due to unsightly scars. On the therapeutic level, the care is multidisciplinary but not consensual, it advocates curbing the flare-ups and treating the lesions formed with the aim of greater comfort for the patient. The objective of this case report is to shed light on this rare disease, on its epidemiological, clinical, and therapeutic aspects, through the study of the management of a case of Verneuil's disease operated on in our plastic surgery department of the Mohamed VI University Hospital in Tangier and through a review of the literature.

Keywords: Verneuil's Disease - Hidradenitis Suppurativa - Apocrine Sweat Glands Multidisciplinary - Surgery.

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INTRODUCTION

Verneuil's disease or hidradenitis suppurativa is a chronic inflammatory and suppurative, fistulizing and sclerosing disease of the pilosebaceous follicles of the anatomical regions rich in apocrine glands. The disease was named by Verneuil, a French surgeon, who in 1864 argued that the initial phenomenon was an inflammation of the sweat glands. This is a rare condition whose incidence has not been clearly established; the prevalence varies from 0.03% to 8% in industrialized countries. The pathophysiology of Verneuil's disease is still subject to much controversy. Its positive diagnosis remains clinical, however there are many differential diagnoses, hence the interest in the para-clinical assessment and in particular the anatomo-pathological study to allow the discernment of these diagnoses.

Although the prognosis is rarely life-threatening, some complications can be serious, both locally (malignant degeneration) and generally, where we note the importance of the psychological and socio-professional impact due to unsightly scars. On the therapeutic level, the care is multidisciplinary but not consensual, it advocates curbing the flare-ups and treating the lesions formed with the aim of greater comfort for the patient.

CASE PRESENTATION

This is a 47-year-old patient, married, originally from and living in Tangier, admitted to us for the management of suspected hidradenitis suppurativa of the left axilla that has been developing for 16 years. On general examination, the patient is conscious and stable in terms of hemodynamics and respiratory function. On

local examination: Diffuse or almost diffuse involvement, and multiple fistulous tracts and interconnected abscesses over the entire left axilla.



Figure 1: hidradenitis suppurativa of the left axilla

Biological tests were carried out and the patient was deemed operable under general anesthesia.

No other paraclinical assessment, including imaging tests, was carried out preoperatively to make a positive diagnosis of Verneuil's disease.

The patient was well informed about the therapeutic strategy with signature of consent.

The goal of surgical treatment was to reduce the extent and progression of the disease, avoid complications and above all improve the quality of life.

The therapeutic strategy was to perform a wide, deep and complete excision of the distribution areas of

the affected apocrine sweat glands of the left armpit in the first surgical stage, then leave the loss of substance in directed healing to take advantage of the retraction, hence the reduction in the diameter of the loss of substance, and then perform a thin mesh skin graft in the second surgical stage.

In the first surgical procedure, the patient underwent a wide excision under general anesthesia, performed in one piece, removing deep skin and subcutaneous cellular tissue, without reaching the musculo-aponeurotic barrier, leaving clear margins. The surgical specimen was sent to anatomopathology for histological study.



Figure 2: wide excision of hidradenitis suppurativa of the left axilla

The defect was left to heal to reduce the diameter of the defect and await the histological results of the surgical specimen.



Figure 3: Secondary healing

In the second surgical stage, the patient underwent a thin graft taken from the inner side of the left thigh to cover the loss of substance in the left armpit

under general anesthesia. The postoperative course was simple with slight lysis of the graft postoperatively without other notable complications.



Figure 4: skin graft after wide excision of hidradenitis suppurativa of the left axilla

DISCUSSION

A. Anatomy of the skin and its appendages

The skin

The different constituents from the surface to the depth of the skin are:

1. The Epidermis

The epidermis, the most superficial layer of the skin, is a keratinized stratified squamous epithelium whose constitution is between 4 different cell populations: keratinocytes, melanocytes, Langerhans cells, Merkel cells.

The epidermis contains no blood or lymphatic vessels, but does contain many free nerve endings.

2. The Dermis

The true framework of the skin, it is made up of fixed cells which are fibroblasts and mobile cells which are blood cells. These cells are associated with collagen, elastin and reticulin fibers. The cohesion of the whole is ensured by the fundamental substance made up essentially of mucopolysaccharides and among them hyaluronic acid. Within the dermis are the vessels which stop at the basal layer of the epidermis.

3. The Hypodermis

The deepest layer of the skin, it constitutes the fat which is more or less thick depending on the individual, it is contained in lobules separated from each other by fibers identical to those of the dermis, these fibers ensuring both the nutrition and the maintenance of the hypodermis.

Skin appendages

1. Sweat Glands

1.1. Apocrine Sweat Glands

a. Embryology

Apocrine sweat glands (or Schiefferdecker glands) are anatomically and embryologically related to sebaceous glands and hair.

Because of this common embryological origin, the apocrine glands are associated with a hair follicle. Their excretory duct opens into the follicular funnel, above the excretory duct of the sebaceous gland.

b. Location

Apocrine glands are distributed mainly in the armpits, ear canals, mammary areolas, pubis, external genitalia. Occasionally, a few isolated apocrine glands can also be found around the navel, on the eyelids, on the scalp, face, and anterolateral surfaces of the trunk.

1.2. Eccrine Sweat Glands

They consist of a sweat gland located in the dermis, which is extended by the excretory duct to the orifice located on the skin surface. These orifices constitute the pores of the skin. Eccrine glands are found on almost the entire integument (2 to 5 million in total, i.e. a density, variable according to the topography, of 150 to 350 per cm²).

2. Hairs

Hair derives from a tubular invagination of the epidermis that extends deep into the dermis.

3. Nails

The nail is made up of 2 parts: A visible part: the body of the nail and a part hidden under a fold of skin: the root.

B. Skin Microbial Flora

General description of the skin flora

Normal human skin is colonized by a large number of organisms that live as commensals on its surface.

We distinguish: A permanent microbial flora and a transient microbial flora.

1. Permanent Microbial Flora

It is a microbial flora residing stably on the skin, most often at the level of the horny layer or at the level of the hair follicle. It is not pathogenic under physiological conditions.

2. Transient Microbial Flora

It is a microbial flora that can temporarily contaminate the skin or settle more permanently in topographies (breeding sites) favorable to humidity and pH conditions (perineum, nasal recesses, external auditory canals) or in the event of a breach of the epidermal barrier.

C. Pathophysiology of Verneuil's disease

The pathophysiology of Verneuil's disease remains unclear at present.

When initially described, hidradenitis suppurativa was considered to be related to inflammation of the sweat glands [1]. Currently, inflammation of the apocrine glands is considered a secondary phenomenon by several authors.

Many studies suggest that follicular occlusion may be the initial phenomenon involved [2, 3]. It would be responsible for a dilation of the pilosebaceous appendix which could lead to its rupture. The discharge of the follicle contents would then cause an inflammatory response with an influx of polymorphonuclear neutrophils, lymphocytes and histiocytes. This inflammatory reaction would lead to the formation of abscesses and then fistulous tracts.

Hidradenitis lesions are coagulase-negative staphylococci and anaerobes. The numerous studies are also contradictory about the possible involvement of these. It seems that bacterial colonization is a secondary phenomenon, potentially responsible for an exacerbation, and not the cause of the disease. Indeed, the culture of aspirate from early lesions of Verneuil's disease is negative in 51% of cases [4].

D. Epidemiological Analysis

Verneuil's disease is a rare condition whose incidence has not been clearly established; the prevalence varies from 0.03% to 8% in industrialized countries according to a study published in June 2015 [5].

Gendre

There is a female predominance with a sex ratio of 1: 2.7 to 1:3.3 according to many studies [1], across the world.

Socio-economic level

1. Occupation

Other studies have revealed a 58.1% absenteeism rate in patients with hidradenitis and that hidradenitis causes a mean loss of 33.6 ± 26.1 working days per year (47). This suggests the significant impact of the disease on work status.

2. Level of Education

In the series by P. Theut *et al.*, [6], patients without a job had a level of education significantly lower than that of patients who remained employed, which

suggests the impact of the disease on the pursuit of studies and hence the reduction in the supply of employment.

E. Clinical Analysis

Risk factors

1. Smoking

Smoking is much more common in patients with HIDRADENITIS compared to the general population. According to a literature review, this association is evaluated with a mean odds ratio of 4.34 (95% CI 2.48-7.60) [7].

2. Obesity

The average body mass index found in our patients is consistent with data from the literature: Overweight is associated with severe forms of HIDRADENITIS [8].

We also note the prevalence of metabolic syndrome which is much higher in patients suffering from HIDRADENITIS than in the general population: between 40 and 50% depending on the studies [16].

3. Genetic

A molecular genetic study found linkage at two loci on chromosomes 6 and 19 in three families, but no linkage of these loci in other families.

Associated diseases

1. Acne

Many publications suggest an association between HIDRADENITIS and acne, this is controversial.

2. Pilonidal Cyst

Pilonidal cysts are common in patients with hidradenitis suppurativa, especially in men (30.2% of patients in the Canoui -Poitrine study [10].

3. Chronic Inflammatory Bowel Diseases

A 9-fold increased risk of developing HPV has been reported in patients with chronic inflammatory bowel disease compared to the general population [11].

4. Neutrophilic Dermatoses

Pyoderma association gangrenosum and HIDRADENITIS is rare but described in several clinical cases and small series.

5. Rheumatological Pathologies

Various rheumatological manifestations have been reported in association with Verneuil's disease. Spondyloarthropathies appear to be associated with HIDRADENITIS: a higher rate of spondyloarthropathy has been reported in patients with Verneuil's disease compared to controls by Richette *et al.*,

Functional signs

1. Initial Reason for Consultation

The rash itself was the primary reason for consultation, followed by the unsightly nature of the scarring, then in equal parts by iterative discharge and pain.

2. Age of Onset of Disease

The average age is usually reported to be 21.8 years in the literature, women seem to develop the disease earlier than men.

3. Delay in Diagnosis of the Disease

The diagnosis of HIDRADENITIS is clinical and does not require any additional examination. The anatomo-pathological analysis of HIDRADENITIS lesions is non-specific and useless for the diagnosis.

Physical signs

1. Topography of Lesions and Number of Affected Areas

The typical distribution of HIDRADENITIS lesions closely corresponds to the anatomical location of the apocrine sweat glands: axillary and inguinal regions. Lesions may also occur in a line connecting these two areas and passing through the breasts and extending to the anal region.

Severity Diagnosis

The severity of HIDRADENITIS is an important parameter taken into account in the reasoning of the therapeutic management of the disease.

1. Hurley Score

The Hurley classification is a simple system for classifying each location into 3 severity grades [12]:

- Hurley I: Abscesses, simple or multiple, without fistulous tract or scar
- Hurley II: Recurrent abscesses with fistula formation and scarring. Single or multiple widely separated lesions.
- Hurley III: Diffuse or near-diffuse involvement, or multiple interconnected fistulous tracts and abscesses over the entire area.

Differential diagnosis

Differential diagnosis is very important in Verneuil's disease given the multitude and its similarity with a large number of diagnoses.

1. Staphylococcal skin infections

2. Epidermal cysts.

3. Scrofuloderma:

This is a cutaneous tuberculosis resulting from damage to the skin by contiguity with a deep tuberculous focus in the lymph nodes or bone.

4. Actinomycosis:

It is a bacterial infection with actinomycetes, in particular actinomyces israelii, a Gram-positive,

anaerobic, endosaprophytic filamentous bacterium of the oral cavity.

5. **Nicolas Favre disease:** This is an infection caused by the L serotypes of chlamydia trachomatis.
6. Crohn's disease
7. Pilonidal sinus
8. Fox Fordyce disease.
9. Acne conglobata
- F. Paraclinical analysis

Bacteriology

Although the etiology of hidradenitis is not known, a wide variety of microorganisms can be isolated from the lesions. The clinical picture of the disease appears to be that of an infectious process and various bacteria are suspected to be responsible for the inflammation.

Imaging

The use of imaging techniques is not widely developed in dermatology, mainly because the skin is immediately accessible for inspection and clinical examination.

Ultrasound and MRI may be instruments of the future for early identification of inflammatory activity of the skin in symptomatic or mild cases because the goal of imaging methods is to make early diagnosis and improve treatment.

G. Therapeutic Analysis

Aim

The Processing is Intended to:

- reduce the extent and progression of the disease
- bring the patient back to an earlier stage
- detect associated diseases
- avoid complications
- treat unsightly scars
- allow psychological support
- improve the quality of life

Means

1. General Measures

1.1. Weight Reduction and Smoking Cessation

There is currently no evidence of the benefit of smoking cessation. However, since tobacco is clearly associated with HS and is generally harmful, stopping smoking is unanimously recommended.

Reducing excess weight is also recommended and appears to be of interest in the management of Verneuil's disease.

1.2. Screening for Associated Diseases

The search for possible associated diseases (IBD, spondyloarthropathy, etc.) should not be neglected, as should the screening of cardiovascular risk factors in order to consider appropriate management.

1.3. Pain Management

Symptomatic treatments such as step I and II analgesics must be combined with specific treatment aimed at controlling the disease.

2. Medical Means

2.1. Local Antibiotic Therapy

Several topical antibiotics are prescribed during Verneuil's disease but the only one to have been the subject of a clinical study is topical clindamycin [13].

2.2. Zinc

Zinc is frequently used when digestive tolerance allows.

2.3. General Antibiotic Therapy

2.3.1. Cyclines

Tetracycline (500 mg x 2/d) was compared to topical clindamycin (0.1% x 2/d) in a randomized trial in patients with Hurley grade I or II hidradenitis suppurativa [14].

2.3.2. Clindamycin – Rifampicin

The use of the clindamycin-rifampicin combination has not been the subject of a comparative study but has been described in 3 open series which each reported complete remission in 8 out of 10 patients [15].

2.4. Retinoids

The presence of follicular occlusion in HIDRADENITIS as in acne, leads to the use of retinoids. Two retrospective studies have not shown any efficacy of isotretinoin, the use of which is currently not recommended for the treatment of HIDRADENITIS [16].

2.5. Biotherapies

2.5.1. Anti-TNF

The use of anti-TNF has been reported in clinical cases and small series and has been the subject of several prospective comparative studies.

Although some results are conflicting, adalimumab and infliximab appear effective for the treatment of severe to moderate hidradenitis suppurativa [16].

2.6. Botulinum Toxin

Botulinum toxin is an experimental treatment for hidradenitis suppurativa. Heeckmann *et al.*, [17], reported that botulinum toxin could decrease sweat production from apocrine glands, in a manner similar to that obtained in eccrine glands.

2.7. Others

Furthermore, corticosteroid therapy (intralesional or systemic), cyclosporine and antiandrogens (cyproterone acetate and estrogens) are reported in several clinical cases or small series. Due to limited data on them, they are not commonly used [16].

3. Physical Treatment

3.1. Laser

In severe to moderate forms, CO2 laser is sometimes used.

Its use aims to destroy all affected tissues by vaporization.

Because of the hypothesis that the disease begins in the hair follicle, Nd-YAG laser hair removal was also evaluated in two studies in which affected areas were treated with 3 to 4 monthly sessions and compared to untreated contralateral areas [18].

3.2. Photodynamic Therapy

Two studies report complete or near-complete regression of lesions after PDT in 4 out of 4 patients [19], while two other studies report no improvement in a total of 10 patients [20].

4. Surgical Means

4.1. Excision of the Lesion

4.1.1. Abscess Drainage

In the face of a painful and abscessed inflammatory lesion, incision/drainage is commonly performed and quickly relieves the patient. This emergency surgical procedure, too often performed on a lesion considered to be a common abscess and without the diagnosis of Verneuil's disease having even been mentioned, is useful if there is indeed a purulent collection; on the contrary, it is useless and even harmful if the nodule is not softened. In all cases, it is a short-term solution, recurrence being generally the rule [21].

4.1.2. Limited Excision

This procedure is useful and better accepted in patients with Hurley stage I, in whom abscess drainage is performed permanently or recurrently.

Limited excision is best done in one piece, removing deep skin and subcutaneous cellular tissue, without reaching the musculoskeletal barrier, leaving clear margins.

4.1.3. Extended Excision

The only radical treatment consists of a wide, deep and complete excision [22], of the distribution areas of the affected apocrine sweat glands both during episodes of superinfections or extensive chronic forms, leaving lateral margins of 1 to 2 centimeters of healthy tissue.

The excision removes deep skin and subcutaneous cellular tissue, without reaching the musculo-aponeurotic barrier.

4.2. Cover Surgery

4.2.1. Direct Suture

This is primary healing, which involves bringing the epidermis and dermis of the two edges of the wound into edge-to-edge contact.

The closure must be done in several layers, each time taking a resistant anatomical structure (aponeurosis, fascia, dermis, etc.); these deep layers help relieve skin tension when the two epidermal edges meet.

Drainage is necessary in the case of a large area of excision, particularly using a Redon drain or Delbey slide.

4.2.2. Directed Healing

a. Principle

It is an alternation of dressings to best direct spontaneous healing.

b. Indication

Its indications are multiple:

- Large wound
- Bruised wound
- Infected wound
- Chronic wound
- Always outside the functional areas, not touching the face, not exposing noble elements, neither tendon nor bone.

It can also be used temporarily, before a skin graft:

- Either to allow a basement to bud and be able to be grafted
- Either to take advantage of the retraction of the bud and thus reduce the surface of the wound before covering it.

c. Epidermization Time

The epidermization period is then approximately six to eight weeks from the surgical creation of the loss of substance.

4.2.3. Reconstruction By Thin or Mesh Skin Grafts

The extent of the excision and the long time for directed healing may lead to proposing the repair of large raw areas, either by skin grafts generally performed in a delayed manner within a period of 2 to 3 weeks or immediately by covering plastic surgery. These techniques ensure faster healing, avoid retractions and vicious scars, but seem to expose to a higher risk of local recurrence [23].

To perform the graft, it is necessary to wait until a quality bud is obtained, non-hemorrhagic and non-infected; the preparation period will require repeated dressings after the excision surgery, a period which can last several weeks, particularly following infectious cellulitis.

a. Principle

This is a thin dermo -epidermal graft whose principle is to leave intact a deep layer of dermis sufficient to contain epithelial enclaves from which an insular epidermization occurs (hair follicles, sebaceous and sweat glands), if the razor does not take keratinocytes, the grafted epidermis does not survive.

A “thin” skin graft must therefore at least pass through the level of the dermal ridges or papillae.

Mesh skin graft aims to multiply the surface area of the graft taken by a variable factor of 1.5 to six (most often three). The epidermalization of the spaces between the meshes of the mesh is done secondarily from the latter.

A mesh graft is secured with a few peripheral stitches or staples, and a few quilting stitches.

b. Post-Transplant

When properly conducted, this technique gives satisfactory results in terms of function and aesthetics; however, it requires a long hospital stay and definitive healing is not generally achieved before 8 to 12 weeks.

Its main drawback remains the little tissue it provides and therefore exposes it to chronic ulcerations [23].

4.2.4. Plastic Reconstruction Using Flaps [24]

a. Principle

It is a fragment of skin, muscle or composite that retains autonomous vascularization by a pedicle. The movements of the flaps are limited by the pedicle.

The use of flaps allows for quick and solid coverage, which is why they are particularly used in noble areas.

However, this involves a longer hospital stay, a potential risk of flap loss due to infection, and a risk of disease recurrence under the flap.

b. Types

- Local flaps
- Regional flaps
- Free flaps (microsurgery)

c. Indications

- Coverage of losses of non-graftable substances (cartilage, tendon, bone, noble elements)
- To shorten the processing time or for aesthetic reasons
- Rotation flaps have generally been used in the inguinal and genital region using surrounding skin.
- If the affected area cannot be completely excised, the use of musculocutaneous flaps

rather than grafts is recommended because of their easier handling in case of recurrence.

5. Psychological Support

By analogy with other chronic diseases, psychological support could be beneficial to patients suffering from hidradenitis and can be offered to them. Patients should also be informed of possible assistance such as off-list ALD and patient associations.

I. Indications

1. Treatment of Hurley Stage I Hidradenitis Suppurativa

In stage I disease, patients frequently have only a few flare-ups per year. Treatment is therefore most often aimed at reducing the duration of flare-ups and may include the following:

- A. Topical treatment: Clindamycin 1% (lotion), Clindamycin 2% (cream), Resorcinol
- B. Short-term systemic treatment: Tetracycline, Erythromycin or other macrolides, Amoxicillin + clavulanic acid – Clindamycin
- C. Azelaic acid, Zinc gluconate

2. Treatment of Hurley Stage II Hidradenitis Suppurativa

Medical treatment aims to control acute inflammatory phenomena and can also be used to prepare the patient for surgery.

Thus the treatment may include:

- a) Medical treatment (systemic only): Clindamycin + rifampicin, Dapsone
- b) Systemic adjuvant therapy or maintenance therapy: Zinc gluconate, Tetracyclines
- c) Surgical treatment: Surgical drainage, Local excision, Laser vaporization

3. Treatment of Chronic Hidradenitis Suppurativa Stage III of Hurley

At this stage, treatment must be medical and surgical.

3.1. Preoperatively

No cure should be expected from medical treatment of stage III disease. All medical treatments are therefore palliative and transient, meaning that the disease reappears shortly after treatment is stopped. The antibiotic combination of clindamycin and rifampicin may also be useful [25].

3.2. Operational Step

In the study by D. Guedes *et al.*, [26], where 20 operated patients underwent a total of 32 surgical procedures in which 54 locations were operated on. On average, they underwent 1.6 procedures and were operated on 2.7 locations.

In the literature, the reconstruction method used varies according to the authors. In a series of 204

operated locations, Bieniek *et al.*, report the use of directed healing (with or without partial suture) for 87 locations (42.7%), thin skin graft for 46 locations (22.6%), direct suture for 51 locations (25%) and the flap for 20 locations (9.8%).

3.3. Post-Operative Stage

3.3.1. In the Medium Term

In the studies of Bordier-Lamy *et al.*, and Alharbi *et al.*, the average length of hospital stay was 6.6 and 5 days, respectively [27]. The average length of hospital stay for grafting was 3.76 days and took place an average of 48 days after excision.

3.3.2. In The Long Term

a. Healing Time

In the literature, the reported healing time is variable: 12.3 weeks for Bordier-Lamy (8.9 weeks after suture or flap, 13.8 after graft and 14.9 after directed healing) [27], and 12.2 weeks after directed healing for Balik *et al.*, [28].

b. Patient Satisfaction

In the study by Bordier-Lamy *et al.*, 32% of women and 9% of men considered surgical procedures “extremely distressing.”

H. Analysis of Evolution and Complications

With treatment

1. Immediate Postoperative Complications

The complications found after surgery for Verneuil's disease in the F. Bordier – Lamy series [27], are: Post-operative infection (11.82%), Suture failure (7.52%), Hemorrhage (2.15%).

2. Post-Operative Complications at a Distance

The complications found after surgery for Verneuil's disease in the F. Bordier – Lamy series [27], are: Persistent pain (18.27%), Lymphedema (2.1%), Fistula (2.1%), Hypertrophic scar (2.1%), Orifice stenosis (1.07%), Graft/flap failure (1.07%), Recurrence (i.e. 2.1%), Bridle (12.9%).

Several articles report lower recurrence rates after wide excision compared to localized excision:

- 27% versus 42.8% according to Ritz *et al.*, after an average follow-up of 72 months [27], (recurrence time of 11 and 20 months respectively on average)
- 0% versus 27% according to Soldin *et al.*, (105)
- 26% versus 38% ($p=0.04$) according to Bordier-Lamy *et al.*, in a French retrospective study of 205 locations operated from 1985 to 2007 (27) (average onset time of 22 months [9-191].

This difference seems to be confirmed by a recent literature review: Mehdizadeh *et al.*, report a recurrence rate of 13% in the case of wide excision, 22% in the case of localized excision and 27% with the

Deroofing technique [30]. However, these data must be analyzed with caution because there is a great heterogeneity between the articles included in this meta-analysis concerning the following data: follow-up time, type of surgery, severity of the disease, locations operated on and method of evaluating the result.

CONCLUSION

It is certain that knowledge about this mysterious and intriguing entity has accumulated. However, it is abundantly clear that many questions remain to be answered. The tools to obtain these answers exist in the modern arsenal of techniques such as molecular biology, imaging, pharmacogenetics, genetics, biochemistry and others. The most delicate of these many questions, which await answers, concern the recognition of early manifestations of the disease that may resemble other unrelated conditions, this early recognition could allow treatments to avoid the progression to the state of terminal debilitating chronicity of the disease for which there is no other therapeutic option than that of a broad surgery.

However, despite all the existing data, it remains difficult to determine which are the best targets for this treatment and when is the best time to perform it.

The recent increase in drug treatment options gives full meaning to the discussion between dermatologist and surgeon. This multidisciplinary approach seems essential to develop in order to consider all possible treatments and to develop a therapeutic strategy adapted to each patient.

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