HIDRADENITIS SUPPURATIVA: A CASE SERIES OF EIGHT PATIENTS

Momčilo STOŠIĆ and Igor STOJANOVIĆ

Summary

Introduction. Hidradenitis suppurativa is a persistent, inflammatory and recurrent disease of the hair follicles which, in time, results in ugly scars. Inflammation and obstruction of channel of sweat glands used to be thought to be the basis of this disease. Case Reports. This paper presents the cases of 8 patients operated on in the past 3 years. A possibility of an oversight in making the diagnosis, as well as an underestimation in the treatment should be taken into consideration. In addition to surgical methods the authors discuss other therapeutic possibilities taken from the available literature. This is a retrospective analysis of the surgical findings. The treatment was multidisciplinary but the surgical excision was the only option which provided healing without recurrence so far. In our series the excision could be done to the fascia or to leave a thin layer of adipose tissue. The reconstruction could be achieved by healing per secondum, by autologous skin or artificial graft, or by narrowing the wound with a skin portion. Our results achieved by stage, secondary stitches and rotation portions were satisfying. Conclusion. In case of advanced disease only surgery can lead to healing. We performed late surgery because the disease was diagnosed in advanced stages according to Hurlay. Wide excision was done in all our patients. The reconstruction was achieved with rotational flap in 3 patients, the wound healing per secondum in 4 patients and with primary suture in one patient. Our patients did not have recurrences of the primary disease. The disease was combined in one case with perianal fistula.

Key words: Hidradenitis Suppurativa; Hair Follicle; Sweat Glands; Surgical Procedures; Operative; Wound Healing

Sažetak


Introduction

In case of hidradenitis suppurativa (HS) most surgeons think that they know what, when and how to treat it. Is that really true? The fact is that there are too many cases when the diagnosis was overlooked [1]. As a matter of fact, it is a common dermatological and surgical problem. Hidradenitis suppurativa is a chronic, recurrent, inflammatory and painful process in the follicles of hair and near a sweat gland. It is characterized by a thickening of the affected skin together with knots and sinuses and development of ugly scars [2]. Topologically, it primarily affects the skin of the folds i.e. the skin of the armpits, groin, genitals and perianal region. The disease was precisely defined at the Second International HS Research Symposium in San Francisco in March 2009 [3]. The synonyms for the disease are acne inversa, Verneuili’s disease and follicular hyperkeratosis [4].

The objective of the study was to show a small series of patients with pictures and definitive results of the surgical procedure. There may have been a possibility of an oversight in making the diagnosis, as well as an underestimation in the treatment. In addition to surgical methods the authors discuss other therapeutic possibilities taken from the available literature.
Case Reports

In the three-year period between 2011 and 2013, 8 patients diagnosed with hidradenitis suppurativa were operated on at the regional Department of Surgery. All the patients were male. The localization in 7 of the patients was the perianal and glutal region. One patient had changes both in the groin and pubis. The age of the patients ranged from 30 to 73 years, the average being 42 years. The retrospective analysis of the surgical findings of all 8 patients, and the interview (with 2 of them after a shorter period of time or a year later) provided the data on the treatment methods applied before the surgery, the length of the treatment, the quality of life before the surgery, especially related to the disease, and, finally, the level of satisfaction after the surgery, both in the functional and aesthetic aspect.

The diagnosis was made in all 8 patients by examination (Figure 1). There was a differential-diagnostic dilemma in 2 patients whether a perianal fistula was present, which proved true in one case intraoperatively. The average length of the treatment before the surgery was over 15 years. They were treated with antibiotics, disinfectants and occasional incisions. Before the arrival to the Department of Surgery, only one patient had an accurately made diagnosis. Two of them were treated with the diagnosis of fistulous sinus pilonidalis, whereas pyoderma and perianal arborized fistula was set as a diagnosis in 3 and 1 patient, respectively. They all had a chronic inflammation of the affected region with a continuous secretion to a larger or smaller extent.

Before the indication was set, the disease had been treated by different specialists (a gynecologist, a general medicine specialist, a dermatologist, a surgeon, an occupational health specialist) as a typical recurrent skin suppuration. The patients had been treated with various antibiotics, dermatological mixtures, occasional incisions, disinfectants and ointments. During that time, scars and channels (sinuses) had multiplied, leaving ugly skin deformities.

All the patients who were surgically treated were at the 3rd stage, according to Hurley. Seven patients were operated under the general anesthesia by the same team, and one received the peridural anesthesia. A wide surgical excision was applied to all of the patients (Figure 2). The excision to the fascia was performed in 5 cases, the excision near the fascia with leaving a layer of fat tissue was done in 3. The injection of methylene-blue into the sinuses was not done. None of the cases included the covering of the tissue defect with a skin graft. The skin defect was narrowed by rotation flaps and Z plastic in 3 patients (Figure 3). A larger part of the postoperative wound healed per secundam (Figure 4).

The patients were interviewed both before and after operations and answered the questions regarding the length of the preoperative treatment and the level of the preoperative dissatisfaction regarding the disease. All the patients had been irritated by the disease manifestation, and two of them claimed it to be a general state of dissatisfaction. One had even been treated for depression.

None of them suffered the HS relapse, except the patient with the perianal fistula (outer opening 10 cm from the anus). The fistula was diagnosed during the operation. All but one patient were operated in the prone position, slightly bent on the operating table. The length of the hospital treatment was between 7 and 14 days or until granulation started to develop, and the total treatment with bandaging in an outpatient department lasted over 2 months, all the way to the complete forming of a stable scar and to a partial epithelization.

During the postoperative interviews, all operated patients showed functional satisfaction, even the one with the fistula recurrence. But the youngest and unmarried patient expressed an aesthetic dissatisfaction. It points to the psycho-social importance of the disease [5].
Stošić M, et al. Hidradenitis Suppurativa

Discussion

No increase in the number of the HS patients has been noticed in the last decades, but it is sure that many papers on the disease have been published in the past 10 years [6]. Although HS is not that rare, both the oversights in making its diagnosis and the underestimation in its treatment are quite frequent. The duration of the disease until the surgery is between 7 and 19 years [3]. As for our patients, the average time elapsed from the onset of symptoms up to the operation was 15 years. The age of all our patients ranged from 15 to 40 years (except the one who was 73), that being the same as in most of the published works. All our patients were male contrary to the statistics found in the literature.

Is it a rare disease or not? By the European criteria, a rare disease is the one with the frequency lower than 5 in 1000 cases or 0.05%, and the American criteria recognize the frequency bellow 0.08%, i.e. less frequently than 1 in 1250 people [7]. The European literature quotes the disease frequency of 1-4% [7, 8]. Some American sources mention the frequency of only 0.053%, which is a greatly underestimated prevalence [9]. The most abundant data are given by Naldi L. in his analysis of 9 studies which mention the frequency ranging from 4 in 100 to 1 in 3000. He explains such a discrepancy by methodological differences. Nevertheless, he comes to a conclusion that the value of 1% is the most precise for many European countries [10]. Most authors quote 2 to 5 times greater frequency in women and the ages affected by the disease ranging from puberty to 40 years of age [1, 11].

Its pathogenesis is unknown. It is common knowledge that hyperkeratotic production occurs in the hair follicle and leads to the occlusion of the output channel. Thus created cystoid formation grows into a small abscess by the intrafollicular bacteria multiplication. The inner follicular pressure increases and causes the rupture of the cystoid formation and the creation of a tract in the surrounding subcutaneous tissues – fistulation. Healing and recurrence of the inflammation lead to the hardening and thickening of the tissue, as well as to the development of the chronic infection with the skin maceration [12]. But the right question is what initiates these processes. Etiological causes are: genetic factors (family predisposition with the frequency of 30-40% [13]), overweight, smoking, changed cutaneo-immunological response and hormone stimulation. The presence and multiplying of bacteria together with a frequent mechanical skin irritation (friction) bring about the development and aggravation of the disease [14]. We think that is the cause of disease in our patients.

Thorough anamnesis and close examination are all we need to diagnose the disease. The localization and characteristic changes strongly suggest HS. A biopsy may be performed only if a combined disease is suspected [3]. The differential diagnosis takes into consideration infected acne, furunculosis, “cat scratch disease”, cellulitis, cutaneous blastomycosis, epidermoid cyst, erysipelas, granuloma inguinale and lym- phogranuloma venereum. It may co-exist with the following diseases: perianal fistula, sinus pilonidalis, Crohn’s disease, irritable colon syndrome, Down’s syndrome, some forms of arthritis, Graves’ disease, Hashimoto’s thyroiditis and Sjögren’s syndrome [12]. HS shares the same immunological etiology with some of these diseases. Certain syndromes, in which HS is a part of the clinical picture, are precisely defined, for example, pyogenic arthritis, pyoderma gangrenosum, acne and hidradenitis suppurativa (PAPASH) – “PASH”, Pyogenic Arthritis, Pyoderma gangrenosum and Acne (PAPA) and others have been noted earlier, too. They are believed to be the consequences of gene mutation [15]. One of our patients had the recurrence of perianal fistula. An internal opening may be found by preoperative physical examination in about 75% of the patients and by transanal ultrasound (TUS) in 95%.

Hidradenitis suppurativa can also be the cause of the squamous-cellular carcinoma, especially in the perianal region [16].

According to the morphological criterion set by Hurley in 1989, the HS classification has 3 stages:
1. Isolated knots (“a blind boil”)
2. Forming of fibroses and fistulous (sinus) channels not connected into a unique area
3. Confluent fistulae, knots and hypertrophic scars [17].

There is also Sartorius’s classification of the HS stages [18]. The treatment mostly depends on the disease stage: mild, medium and heavy forms which correlate with Hurley’s morphological picture. Even a few lethal outcomes due to HS have been reported. Hidradenitis suppurativa was classified into Hurley stage III in our patients. The quality of life is always severely decreased in the HS patients, especially in the stages of the disease which are graver and more spread on the skin. In the most frequently affected age group, the disease can cause both psychological and physical disorder of the patients’ sex life. In any case, the social life of the patients is affected, and their professional abilities are diminished. That makes the social support and psychological treatment necessary.

The basic principle in the management of HS is that an early and thorough surgical excision gives the best results. The treatment includes:
– a local treatment: disinfectants, light (photodynamic), laser treatment, local antibiotics and other solutions, which are not used excessively;
– a pharmacological and medicamentous treatment: immunosuppressants and immunomodulators, tumor necrosis factor (TNF) alpha inhibitors, antibiotics, oral retinoids and anti-androgens, which are not at our disposal;
– a surgical treatment: “deroofing”, wide excision; skin grafts and other esthetic operations as a temporary solution;
– a radiological treatment – an external radiation, not used by us;
– and a psychological treatment.

A radical surgery can be delayed in stage I. As for the later stages, it is best to have a surgical intervention as soon as possible, as our experience shows. Generally, the operation can be delayed during stage I. Instead, it is recommended to shave the region and administer antibiotics according to the antibiogram. Local treatment of rinsing with antibiotic and disinfecting solutions is also recommended. The most common isolated causes of an infection are: Staphylococcus aureus (51%), followed by Escherischia coli, Streptococcus, and anaerobes [15]. Various antibiotics can be administered, such as clindamycin, cephalosporin, tetracycline, rifampicin and others. Photodynamic therapy has been introduced recently, as well as laser therapy for stages I and II [15]. However, neither of these two therapies has been used in our cases. The change of lifestyle in terms of a weight loss and giving up smoking affects the treatment results in some patients. Because of the high percentage of recurrence, the patient should be prepared psychologically for a long-lasting treatment and, finally, a surgical intervention [17].

According to Hurley, the surgical treatment is applied in stage II and III. It can be performed alone or in the combination with a medication therapy.

Available literature data show that the following medications are used in the treatment: immunomodulatory medications (anakinra, cinakinumab, and ustekinumab), which improves the condition in 2/3 of the HS patients and is also used in psoriasis, as well as azathioprine, tacrolimus, thalidomide) [7], anti-inflammatory TNFα-inhibitors (infliximab, etanercept, adalimumab), immunosuppressors (prednisone), dapson, antiandrogens and acitretin. They have good effects when the disease is of the autoimmune genesis. Since all our patients were in stage III of the disease, this treatment was not used.

Surgical treatment of our patients always included a wide excision of the change in the gluteal region going to the fascia in 5 cases (one of them with several different excisions of the affected cutaneous and subcutaneous areas) and with the removal of the most part of the subcutaneous, fat tissue except a thin layer above the fascia i.e. “almost to the fascia” in the other 3 cases. Most authors agree that the surgical treatment has no alternative in other types of treatments if we want to come to a definite solution of the problem [19–22]. A dilemma is whether to perform the excision to the fascia or “almost to the fascia” until the granulation begins [23]. At the same time we had in mind the principles of plastic surgery [24].

After the excision of the affected region, the following is possible: 1. leaving the wound to heal per secundam, 2. healing per secundam with a partial closing of some of the portions, 3. delayed grafting with an autologous skin portion in its full thickness [23], and 4. delayed grafting with an artificial graft [25]. The choice depends on the size of the defect, risk of a recurrence, risk of an infection and expected aesthetic result. The healing per secundam and partial narrowing of the excised area by skin portions, Z plastic, etc. have the lowest percentage of recurrence and infection but the longest treatment and poorest aesthetic result [25]. The grafting with skin in its full thickness gives good results, but has a higher percentage of infection [26]. More and more authors advocate the grafting with an artificial skin graft. However, considering the graft thickness and aesthetic aspect, it is not recommended to do the excision to the fascia, but more moderately [27].

We did not perform the surgical procedure of cutting and opening the sinus tracts with the removal of their surface parts (“deroofing”) and we think it is a palliative measure with a high percentage of relapsing [28]. It is good for the management with multiple suppurations in an outpatient department. We used simple and multiple incisions as a temporary procedure.

Some time ago, many authors recommended X-rays and laser to be applied before the final operation (neodymium: yttrium-aluminium-garnet laser to be applied for three months or carbon dioxide laser). The logical goal was to “dry” the affected area by the application of the outer radiation in the amount of 3–8 Gy in several fractions. A study performed in 2000 reported radical withdrawal of the changes in 1/3 of 231 cases [22].

Conclusion

Hidradenitis suppurativa is not a rare disease. The choice of the treatment methods is wide. It is sure that keeping records of the patients improves the treatment – as they do in the Scandinavian countries with the so-called HISREG (Clinical Scandinavian Registry for Hidradenitis Suppurativa) method of case registration. According to our experience, surgical treatment is not indicated in the early stages of hidradenitis suppurativa. In the advanced stages of disease, only a surgery can lead to healing. The affected regions are characteristic. The affected region in all our patients was the gluteal region. When scars and sinus fistulae begin to form, an early operation prevents the creation of more serious esthetic consequences. The operative management is a wide excision without compromise and with one of the described ways of the operative wound closing. Recurrences on some parts of the
operated area are possible even after the surgery. In that case, a re-excision is performed. Our patients did not suffer recurrences of the primary disease.

The disease may be combined with other diseases such as: fistulae, pilonidal sinus, immunosuppressive states, or within the mentioned syndromes.

References

23. Marzano AV, Trevisan V, Gattorno M, Ceccherini I, De Simone C, Crosti C. Pyogenic arthritis, pyoderma gangreno-