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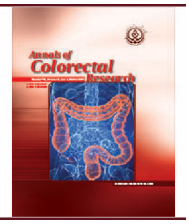

Severe Perineal Hidradenitis Suppurativa and Fistulas: An Underrated Association?

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Abstract

Background: Due to its multiple forms of presentation, hidradenitis suppurativa has classically imposed a diagnostic challenge. The prevalence of perianal fistula in patients with hidradenitis suppurativa ranges from 6.6% to 67%. The aim of the study was to assess both conditions.
Methods: A retrospective chart review of the years 2000 to 2018 was conducted for patients with hidradenitis suppurativa using the ICD-9-CM code 705.83. Hurley's three-stage classification was applied. Diagnosis and relevant patient characteristics were assessed, along with the presence of associated perianal fistula. Endoanal ultrasound (EAU) had been performed in 61% of the patients with perianal hidradenitis and perianal fistula, whereas magnetic resonance imaging (MRI) had been done for 19%.
Results: Of 143 cases with hidradenitis, a perianal (perineal/buttocks) presentation occurred in 62 (43.4%) of patients, among whom 93.5% were men. Twenty-one percent were associated with perianal fistulas, six being complex ones associated with Hurley stage II and III. Treatment for the latter included: loose setons in four patients with Crohn's disease and two non-Crohn's disease with complex fistulas; four fistulotomies and two fistulectomies in low transsphincteric fistulas; and one incision and drainage.
Conclusion: Perianal fistula should be treated according to associated diseases and the type of fistula. Association of hidradenitis and perianal fistulas may be higher than expected and the relation of severe hidradenitis with complex perianal fistulas should be studied further. Endoanal ultrasound and MRI may be useful tools to assess HS with complex perianal fistulas, but the iconographic patterns of hidradenitis and Crohn's disease should be kept in mind as an association may prevail.

Keywords: Hidradenitis suppurativa, anal fistula, Crohn's disease, diagnosis of hidradenitis suppurativa

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Introduction

Hidradenitis suppurativa (HS) has been a troubling entity for the general surgeon due to the high percentage of recurrences (over 30%) and unsatisfactory surgical interventions, especially in severe cases. The increased interest in HS seems strongly related to the 2005 approval of the monoclonal

antibody, Adalimumab, in Europe for the treatment of severe HS, although other treatments have also been applied (1). In Spain, this has specifically led to the endorsement of the Hercules project by twenty scientific societies, including the Spanish Association of Surgeons, to establish a diagnostic and therapeutic algorithm for HS. This has helped to assess the estimated prevalence of HS, which involves over 1% of the general population (2). Prevalence of HS in patients with perianal fistula (PF) ranges between 6.6 to 64% (3, 4) and should be part of the differential diagnosis of HS avoiding undiagnosed PF. Therefore, the presence of anal fistula, may be as well, the first expression of an underdiagnosed HS. Recurrences are common and highest in perianal HS compared with other body sites (5). Perianal HS can coexist with other inflammatory disorders such as Crohn's disease, complicating their management (6). The aim of this study was to assess HS and its association with PF.

Methods

A retrospective chart review of the years 2000 to 2018 was conducted using the ICD-9-CM (International Classification of Diseases) code 705.83 for patients with HS. These patients were primarily seen by surgeons and there was no specific protocolized treatment other than surgery. Severity was established according to the three-stages of Hurley's classification (7). Diagnosis and relevant patient characteristics were assessed, as was the presence of associated PF. A recurrence was defined as symptoms recurring within one year after the last treatment. The number of surgical procedures each patient underwent was also noted. Data collection was done until the last follow-up visit. Imaging was not routinely used, but PFs had been assessed by bi-dimensional endoanal ultrasound (EAU) using a B-K Medical Falcon 2101 ELX, available from January 2006 onwards. On the other hand, MRI had been performed only for severe HS using a Philips Ingenia 1.5 Tesla device. No skin ultrasounds had been performed. We tried to assess retrospectively the need for multidisciplinary approaches. From 2018 onward, patients with HS were referred first to a dermatologist in our hospital.

Statistical analysis was performed with the SPSS 12.0 package for Windows. The quantitative variables were expressed as mean with standard deviation. Qualitative variables were expressed in percentages.

Results

One-hundred and fifty-four patients matched the ICD-9-CM code 705.83 for HS. Eleven cases were excluded for inconsistent HS diagnosis, meaning that 143 patients were included in the study. Main differential diagnosis was with pilonidal disease and perianal abscess (Figure 1). The characteristics of HS are listed in Table 1. Hurley stage III was diagnosed

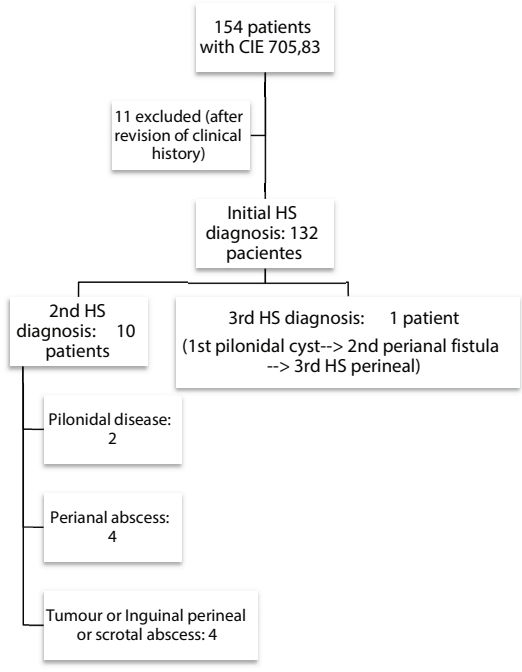

Figure 1: Total number of HS and differential diagnosis

Table 1: General characteristics

Total number of patients	143
Men	86 (60.1%)
Women	57 (39.9%)
Mean age and SD*	39±12
Comorbilities of HS	
Tobacco	72 (50.3%)
Obesity	21 (14.7%)
Anxiety/depression	49 (34.3%)
Crohn disease	4 (2.8%)
Psoriasis	12 (8.4%)
Other autoimmune diseases	5 (3.5%)
Alcohol abuse	8 (5.6%)
General Location	
Axilar	34 (23.8%)
Axilar + Inguinal	2 (1.4%)
Axilar + Perianal	1 (0.7%)
Inframamary	1 (0.7%)
Inguinal	41 (28.7%)
Inguinal + Axilar	1 (1%)
Inguinal + Axilar + Genital	1 (0.7%)
Inguinal + Perianal	10 (7%)
Perianal	51 (35.7%)
Other	2 (1.4%)
Perianal (perineal/buttocks) location	62 (43.4%)
Men	58 (93.5%)
Follow-up	46 (74.2%)
Operated	61 (98.4%)
Recurrences	31 (50.8%)
At least second surgery due to recurrence	19 (61.3%)
Plastic surgery	8 (13%)
Dermatology	13 (21%)
Perianal Fistula	13 (21%)** (9.1% from 143 patients)

*SD= Standard deviation ** Characteristics of PF on Table 2

Table 2: Perianal fistula characteristics

Crohn's disease	Gender	Age	HS localization	Hurley grade	Type of PF*	EAU**	MRI***	Surgery	Reintervention	Follow-up
Yes	Male	46	Scrotal	3	Complex	1st: Transphincteric high 2nd: Suprasphincteric	No	Seton	Seton	Yes
Yes****	Male	33	Perineal	3	Complex	Transhincteric high	Low	Seton	3 seton changes +1 fistulotomy	Yes
Yes****	Male	40	Perineal	2	Complex	No	High	Seton	No	Yes+
Yes	Male	53	Inguinalscrotal	3	Complex	Transhincteric high	No	Seton	Seton	Yes
No	Male	38	Perineal	3	Transphincteric low	No	No	F-lectomy	No	Yes+
No	Male	46	Inguinalperineal	1	Transphincteric low	No	No	F-lectomy	F-lotomy	Yes
No	Male	51	Perineal	3	Complex	Transphincteric high + Intersphincteric	No	Seton	Seton change	Yes
No****	Male	35	Perineal	2	Transphincteric low	Subcutaneous tract	No	F-lectomy	No	No (Lost)
No	Male	48	Gluteal	3	Complex	Transphincteric high + low	High + Low	F-lotomy + partial F-lectomy	Seton	Yes
No	Male	26	Perineal	1	Transphincteric low	Low	No	F-lotomy	Incision + drainage	No (Lost)
No	Male	47	Perineal	1	Intersphicteric low	Low (x2)	No	F-lotomy	F-lotomy	Yes
No	Male	32	Perineal	1	Transphincteric low	No clear fistulous tract	No	F-lotomy	Nodule excision	Yes
No	Male	24	Inguinal	1	Transphincteric low	Low	No	Partial F-lectomy	F-lotomy + Seton	Yes

* Type of perianal fistula ** EUA Endoanal ultrasopund, *** MRI magnetic resonance image, **** Psoriasis +Other hospital, All seton placed were loose setons

Figure 2: Hurley's stage disease

in 20% of the series (Figure 2).

The presentation was in perianal (perineal/buttocks) location in 62 patients (43.4%), most of whom were men (93.5%). Notably, EAU was conducted for 69% of patients with HS and PF, while MRI was done for 23%.

Follow-up was possible for 46 patients (74.2%) with HS in the perianal (perineal/buttocks) location, as described in Table 1. The recurrences after the first surgery were 50.8%, while 61% required at least another surgery. A multidisciplinary approach was taken in a total of 21 patients (34%); the plastic surgery specialists intervened in eight severe HS cases, while the dermatologists treated 13 cases with recurrences or other associated dermatologic diseases (Table 1).

Thirteen patients (9.1%) had an associated PF treated with (last surgery): setons in four patients with Crohn's disease and two non-Crohn's disease patients with complex fistulas; and four fistulotomies and two fistulectomies in low transsphincteric fistulas. Transsphincteric and suprasphincteric fistulas were defined as complex fistulas, which had been mainly treated with loose setons. Complex fistulas were more associated with Hurley's stage II and III. Two cases were lost on follow-up and two made their follow-up in their reference hospital. Table 2 delivers type of fistula, EAU, MRI, surgery, follow-up and Hurley's stage.

Discussion

Hidradenitis suppurativa is a chronic, auto-inflammatory skin disease. Due to its multiple forms of presentation, HS has been a diagnostic challenge. The rate of diagnostic error in our series over an 18-year period was 7.7% (8). Especially in the early stages, it can be confounded with other entities such as pilonidal sinus, cellulite or perirectal abscess; in more advanced stages, it is difficult to distinguish from dermopathies, such as psoriasis (12 cases in our series) and classic acne.

In recent years, dermatologists have included skin ultrasound to complement the diagnosis and establish the prognosis of this entity. In 2015, Martorell et al. (9) described the tendency to underestimate the

severity of the lesions when only taking into account the clinical assessment, proposing the regular use of ultrasound of the skin for detection of the subclinical disease. This would allow a more real staging, as well as a dynamic monitoring of the inflammatory activity of the disease, improving its management.

In another study conducted by Wortsman et al. (10), ultrasounds performed on 34 patients with a total of 142 HS lesions allowed the determination of subclinical collections in 76%, fistulae in 29%, and dermal pseudocysts in 71% of cases. These ultrasound findings led to a change in clinical treatment in 82% of cases; in 24% of cases, medical treatment was switched to surgical management.

In the particular case of severe perianal HS, the association with PF shows a wide range according to different publications, with a minimum of 6.6% in a recent published series (4). In fact, the performance of EAU and transperineal ultrasound in severe perianal HS may objectify subclinical PF that is later confirmed in the operating room (11), showing that the association of severe perineal HS and PF might be higher. In our series, in 13/62 patients with perianal (perineal/buttocks) involvement who also had PF, four had Crohn's disease but nine did not. There may be a trend to relate a higher Hurley stage with a complex PF, but four such patients had Crohn´s disease and the number of patients is too small for such a statement (6 out of 13).

In this series, MRI had only been performed when complex fistula was suspected (HS not included in a protocolized study). The follow-up of two patients took place in their reference hospital. Our approach to PF is conservative although a modified core out technique for anal fistula was described previously (12). Subsequently to the introduction of the Hercules project in 2018, HS patients have initially been seen by dermatologists before being referred for surgery if needed. We could show that a multidisciplinary approach was needed in 21 patients (34.1%) with more severe disease in the perianal location.

In the presence of anal fistulas and suspected HS, MRI is also recommended to define the extent of the disease and to exclude perianal Crohn's disease, with which HS may also be associated, as well as other autoimmune and skin conditions. Although, HS and Crohn's disease may coincide, following Monnier et al. (13), the specific diagnosis by MRI in the case of HS is possible with a combination of three basic items: lower presence of fistulas and sphincter involvement, absence of thickening of the rectal wall and bilateral lesions. Griffin et al. (14) evaluated 31 MRI's performed in 18 patients with anogenital HS. Most patients were men and only three cases had synchronous Crohn's disease. The main involvement was from a large but shallow region, scarcely affecting the anal sphincter. In the case of recurrent perianal HS, Derruau S et al. (15) propose the intraoperative use of Infrared Thermography in combination with preoperative MRI to ensure an adequate margin of healthy tissue during the excision, thereby reducing the risk of recurrence.

Although HS is three times more frequent in women, perianal (perineal/buttocks) HS involvement occurs mostly in men (93.5% of our series) and is associated with obesity and smoking.

The standard treatment of HS is antibiotic therapy and drainage of abscesses with subsequent cures. Notably, anti-TNF (tumor necrosis factor) drugs used by dermatologists seem to improve the patient's clinical status in the face of surgical intervention, decreasing inflammation (2). In fact, the current interest in HS with PF has led even dermatologists to publish a therapeutic algorithm for HS and anal fistulas (16). They propose the excision of the most superficial cutaneous tracts and deroofing with healing by second intention of the deep ones. In the case of anal fistulae, they propose fistulotomies in simple ones and the placement of a loose seton with a second surgery in complex fistulas. The commonly performed treatment for perianal Crohn's disease is mainly by a draining seton (70.7% of cases), as in our series, although other standard techniques are quoted by Lamb et al. (17). Fistulas in non-Crohn disease patients should be treated following the recommendations of Williams et al. (18). The possibility of using stem-cell therapy for the management of complex PF in order to reduce the risk of fecal incontinence should also be mentioned. Recently, the application of stem cell therapy on 52 anal fistula cases with a 48-month follow-up period was reported. Eighteen cases were Crohn's disease and one was HS. Since 2018, allogenic stem cells (Alofisel®) are available and authorized by the European Medicine Agency (EMA), with a 55.5% healing rate being achieved in Crohn's disease (19).

The limitations of this study included its retrospective nature and minimal data regarding follow-ups.

Conclusion

Perianal fistulas should be treated according to the associated diseases and type of fistula. Association of hidradenitis and PF may be higher than expected, and the relation of severe hidradenitis with complex PFs should be studied further. Endoanal ultrasound and MRI may be useful tools to assess HS with complex PFs, but the iconographic patterns of hidradenitis and Crohn's disease should be kept in mind as both may be associated.

Conflicts of interests: None declared.

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