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Differential Diagnosis Between Perianal Crohn's Disease and Hidradenitis Suppurativa: A Challenging Teamwork

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Abstract: Hidradenitis suppurativa (HS) is a rare, debilitating skin disease characterized by the presence of recurrent tender subcutaneous nodules that develop into abscesses and fistulae. Isolated perineal Crohn's disease (CD) is unusual, diagnosis can be difficult, and distinction from HS is a challenge for the gastroenterologist. The aim of this work was to determine the criteria that distinguish perineal CD from perineal HS. Four patients with isolated perineal CD and three with perineal HS were included. Rectal or skin biopsies of all CD patients showed granulomas. No granulomas were found for HS. Fistulae were present in 4/4 CD, extended to the anal canal. All patients with HS had gluteal abscesses. They were bilateral in all cases, superficial. Perineal lesions management should involve a multidisciplinary approach in order to make an accurate diagnosis and ultimately to give the best and most effective treatment.

Key Words: Crohn's disease, Hidradenitis suppurativa

What Is Known

- Hidradenitis suppurativa (HS) is a rare, debilitating skin disease characterized by the presence of recurrent tender subcutaneous nodules that develop into abscesses and fistulae.
- Isolated perineal Crohn's disease (CD) is unusual, diagnosis can be difficult, and distinction from HS is a challenge for the gastroenterologist.

What Is New

- MRI presentation of anoperineal disease may overlap between CD and HS. But, diagnosis of HS is possible with a combination of 3 features: either posterior localization of the lesions or absence of features' predominance in perianal area, absence of rectal wall thickening, and bilaterality of features. HS lesions in MRI are superficial.
- MRI with skin or rectal biopsy could be the more contributory complementary examination.

INTRODUCTION

Hidradenitis suppurativa (HS) is a rare, debilitating skin disease characterized by the presence of recurrent tender subcutaneous nodules that develop into abscesses and fistulae. Lesions may develop in the intertriginous apocrine gland-bearing areas of the body, in decreasing order of frequency axillary, inguinal, perianal, perineal, mammary, buttock, pubic, chest, scalp, retroauricular, and eyelid, usually after puberty (1, 2).

The diagnosis of HS is primarily made based on its characteristic clinical presentation and has to meet the criteria adopted by the 2nd International Conference on HS, March 5, 2009, San Francisco, CA. All three criteria must be met for establishing the diagnosis (3):

(1) Typical lesions, ie, deep-seated painful nodules: "blind boils" in early lesions; abscesses, draining sinus, bridged scars, and "tombstone" double-ended pseudocomedones in secondary lesions.

(2) Typical topography, ie, axillae, groins, perineal and perianal region, buttocks, infra, and inter mammary folds.

(3) Chronicity and recurrences.

The severity of the disease can be classified in 3 grades for each area involved according to the Hurley classification (4).

HS has an estimated prevalence of 1%–4%, with a notable female-to-male predominance of 3:1 (1, 2).

Crohn's disease (CD) is a chronic inflammatory bowel disease that is characterized by segmental noncaseating granulomas and may affect any portion of the gastrointestinal tract from mouth to the anus.

Perineal CD presents as fistulas and fissures, associated abscesses, uncomplicated skin inflammation, and skin tags (5).

Isolated perineal CD is unusual, diagnosis can be difficult, and distinction from HS is a challenge for the gastroenterologist (6–8).

The aim of this work was to determine the criteria that distinguish perineal CD from perineal HS.

MATERIAL AND METHODS

All patients with a diagnosis of isolated perineal CD according to the European Society of Pediatric Gastroenterology and Nutrition criteria (9) or of perineal HS followed in our tertiary center were considered for inclusion in this retrospective study. Data were retrospectively obtained from medical files and recorded in an anonymous standardized form. The following baseline characteristics were recorded for each patient: gender, age at onset, weight, body mass index, anti-*Saccharomyces cerevisiae* antibodies (ASCA), MRI, biopsies, extraperineal locations, CD classification according to Paris classification, and HS according to Hurley classification.

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TABLE 1. Clinical characteristics at diagnosis of isolated perineal Crohn's disease and hidradenitis suppurativa

Characteristics	CD1	CD2	CD3	CD4	HS1	HS2	HS3
Gender	Girl	Girl	Girl	Boy	Boy	Boy	Boy
Age at diagnosis	15	9	10	16	15	15	9
Family history	None	None	None	None	None	Father similar lesions nape and chest without diagnosis	None
Symptoms duration (mo)	36	1	4	3	14	4	36
BMI	18	20	15	17	22	20	20
Perineal lesions	Fistulae	Fistulae	Fistulae	Fistulae	Abscess	Abscess gluteal	Abscess Gluteal
					Natal cleft	Bilateral	Bilateral
Extraperineal locations	Height delay	Aphtae	Height delay	None	Chest	Nape	None
Paris classification	A1b	A1a	A1b	A1b			
	B3p	B3p	B3p	B3p			
	G1	G0	G1	G0			
Hurley classification					I	III	II
Treatment	Azathioprine and infliximab	Azathioprine and infliximab	Infliximab	Azathioprine and infliximab	Antibiotics infliximab	Infliximab antibiotics	Adalimumab antibiotics

BMI = body mass index; CD = Crohn's disease; HS = Hidradenitis suppurativa.

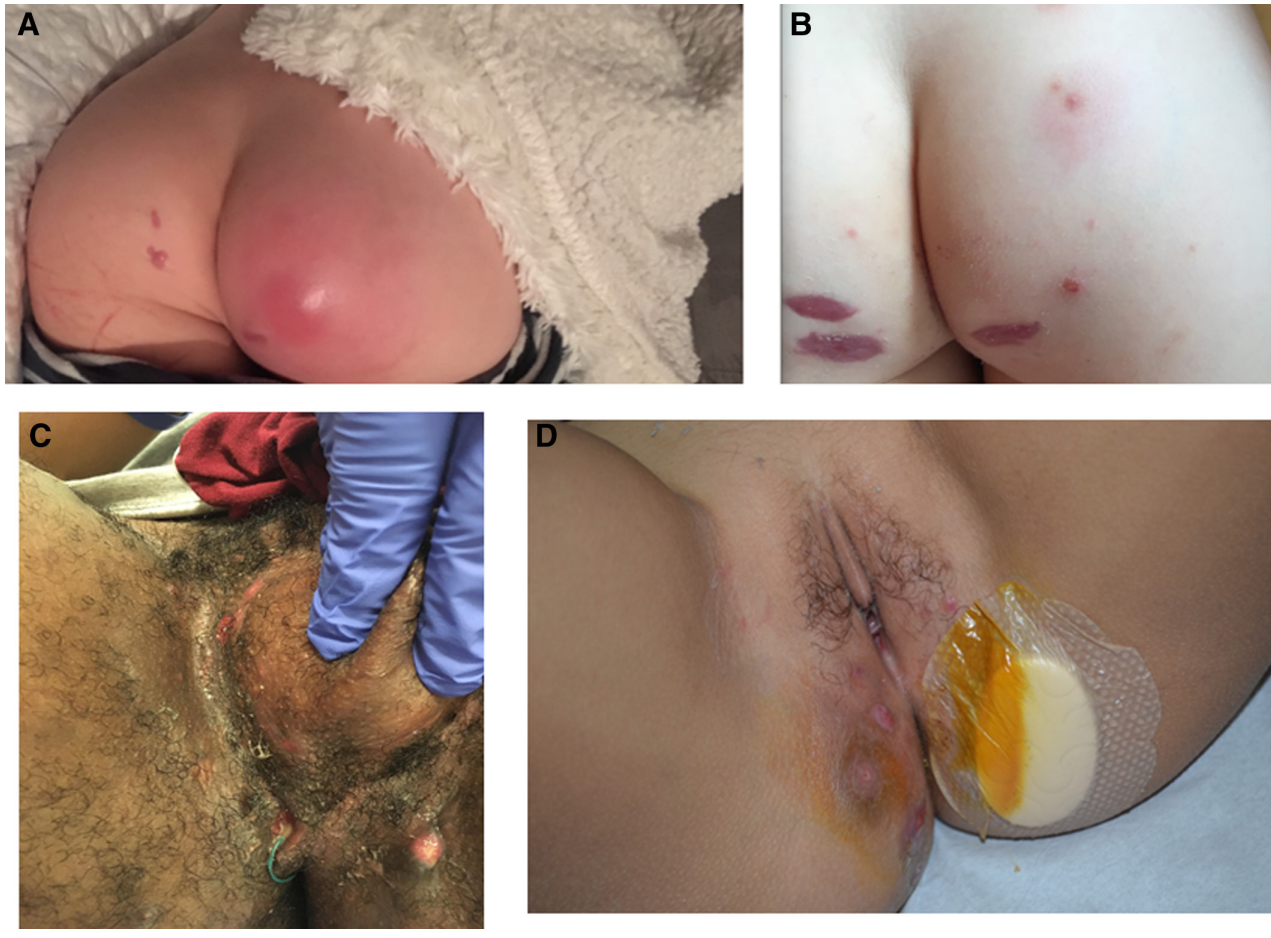

FIGURE 1. Gluteal abscess before (A) and after (B) surgery, Hurley I, in a 9-year-old boy. C) Perineal abscesses, Hurley III, in a 15-year-old boy. D) Perineal Crohn's disease in a 10-year-old girl.

Patients with luminal CD were excluded (normal colonoscopy or normal calprotectin).

RESULTS

Four patients with isolated perineal CD and 3 with perineal HS were included.

Clinical data are summarized in Table 1, and pictures shown in Figure 1.

Biopsy

Among CD patients, granulomas were found on skin biopsies (n = 2) or rectal biopsies (n = 2). No granulomas were found in HS patients.

ASCA

Three (of 4) CD patients were ASCA+. This data was not available for HS patients.

MRI Features

Morphological findings in patients with HS and CD are listed in Table 2, shown in Figure 2.

Fistulae were present in 4/4 CD. Fistulae in CD extended to the anal canal with no penetration into the sphincter mechanism. Location in CD was perianal.

TABLE 2. Pelvic MRI findings in patients with hidradenitis suppurativa and Crohn's disease

Morphologic characteristics	CD	HS
Inflammatory features		
Abscess	0/4	3/3
Sinus tract	4/4	1/3
Classification		
Complex	0/4	0/3
Intersphincteric	0/4	0/3
Extrasphincteric	4/4	0/3
Transphincteric	0/4	0/3
Suprasphincteric	0/4	0/3
Superficial	0/4	3/3
Penetration into the sphincter mechanism	0/4	0/3
Anatomical regions		
Ischiorectal fosse	0/4	0/3
Sacral/gluteal	1/4	3/3
Anterior/inguinal	0/4	0/3
Natal cleft	2/4	0/3
Perianal	4/4	0/3
Additional features		
Lymphadenopathy	3/4	0/3
Rectal wall thickening	1/4	0/3
Myositis	0/4	0/3
Subcutaneous edema	2/4	2/3
Skin thickening	1/4	1/3
Global features		
Predominance in perianal area	3/4	0/3
Bilaterality	1/4	3/3

CD = Crohn's disease; HS = Hidradenitis suppurativa.

All patients with HS had gluteal abscesses. They were bilateral in all cases, superficial, in one case extended to the sacral region.

Treatment

All CD patients were treated with infliximab, 3 patients received azathioprine.

All HS patients were treated with antibiotics. One HS patient improved on infliximab and another one on adalimumab. The third patient worsened on infliximab and adalimumab and required intravenous antibiotic treatment and surgery.

DISCUSSION

Only 4 patients with isolated perineal CD were identified among the 484 CD followed at our tertiary center (0.82%), whereas 15.7% (n = 76) had perianal and luminal disease. Isolated perineal CD is , and there may be confusion with other perineal lesions such as HS. Both diseases are rare; thus, we only can present a small series of patients.

HS and CD share many characteristics, and the two diseases can be associated in the same patient (7). The prevalence of HS in CD patients is 12.4%–17.9%, while the prevalence of CD in HS patients is around 3%.

Family history and extraperineal skin locations can help to guide the diagnosis. Physicians should inspect the axilla, chest, inguinal creases, vulva, perineum, perianal skin, buttocks, and medial thighs for other signs guiding to HS such as comedones, acneiform papules and pustules, inflammatory nodules, sinus tracts, ulcers, erosions, drainage, granulation tissue, induration, edema, lymphedema, lichenification, dyspigmentation, scars (atrophic, hypertrophic, keloidal), strictures, and contractures (10,11).

Biopsy is warranted if CD is suspected. A biopsy of the affected skin showing granulomatous inflammation on histopathology strongly supports a diagnosis of CD. Sometimes granulomas are found only on rectal biopsies suggesting that a colonoscopy should be performed for an accurate diagnosis (12).

ASCA could help to diagnose CD. Some studies provided preliminary evidence, that inflammatory bowel disease may have a preclinical phase that might be detected by disease-specific biomarkers, such as ASCA. These biomarkers should be part of the diagnosis process (13). A recent study (14) identified serologic detection of ASCAs as a biomarker for HS, mainly in its severe (Hurley III) and inflammatory forms. The association between ASCA seropositivity and high HS tissue and/or systemic inflammatory burden provides support for a link between systemic inflammation in the absence of inflammatory bowel disease and the development of ASCAs. It has been showed in CD that positivity for ASCAs is associated with more severe disease and greater need for surgery (15).

MRI presentation of anoperineal disease may overlap between CD and HS. But, diagnosis of HS is possible with a combination of 3 features, as already shown by Monnier et al (16): either posterior localization of the lesions or absence of features' predominance in perianal area, absence of rectal wall thickening, and bilaterality of features. HS lesions in MRI are superficial. These points should be discussed with the radiologist to guide the diagnosis (16,17), and this may be with biopsy the more contributory complementary examination.

Medical treatment of perineal disease in CD includes antibiotics, anti-tumour necrosis factor (TNF) antibodies. In HS, multimodal treatment should be discussed by gastroenterologist and dermatologists. The choice of therapy is guided by disease severity (18). Antibiotics are widely used, from oral to intravenous route. Patients with moderate to severe disease can also be treated with anti–TNF-α antibodies, especially if they have required long-term antibiotic treatments. Adalimumab was approved for the treatment of HS by the

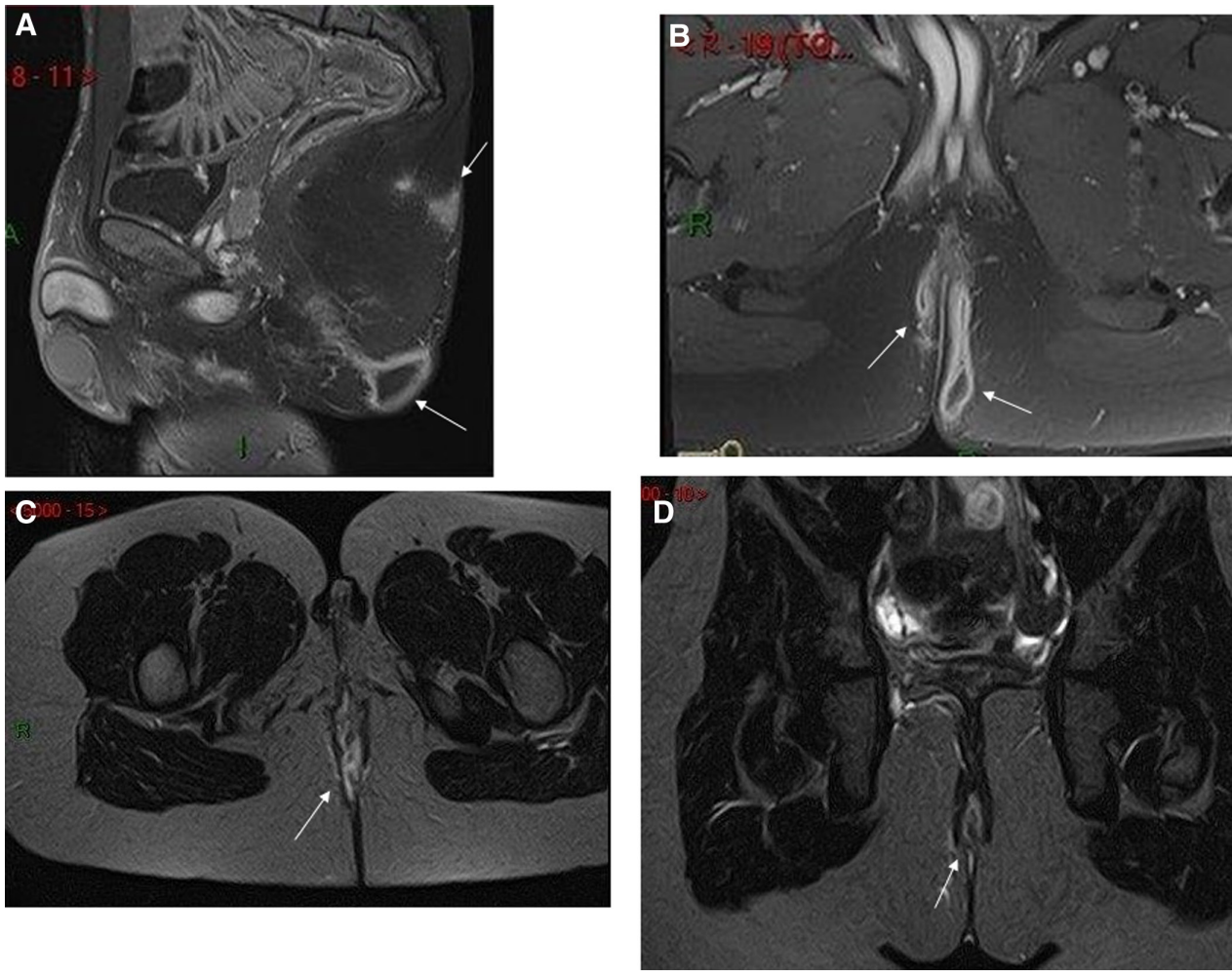

FIGURE 2. A) Fat suppressed gadolinium enhanced sagittal T1 weighted MRI in a 15-year-old boy with perineal hidradenitis shows a posterior gluteal abscess with sacral subcutaneous oedema. B) Fat suppressed gadolinium enhanced coronal T1 weighted MRI in a 15-year-old boy with perineal hidradenitis shows a posterior bilateral gluteal abscess. C) Fat suppressed axial T2 weighted MRI in a 9-year-old girl with perineal Crohn's disease show a posterior median fistula. D) Fat suppressed coronal T2 weighted MRI in a 9-year-old girl with perineal Crohn's disease show a posterior median fistula.

US Food and Drug Administration in 2016 (19). Infliximab may be used as an alternative anti-TNF therapy. In case of treatment failure, third-line should be discussed (anakinra, ustekinumab, dapsone, or acitretin) (20). Surgery is required to definitively treat the tunnels and scars associated with chronic HS.

In conclusion, management of perineal lesions should involve a multidisciplinary approach combining the knowledge of the gastroenterologist, the dermatologist, and the radiologist who have appropriate experience in this area in order to make an accurate diagnosis and ultimately to give the best and most effective treatment.

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