Hidradenitis suppurativa and Crohn's disease: Two cases that support an association

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ABSTRACT

An association between Crohn's disease and hidradenitis suppurativa has been suggested. The presence of Crohn's disease generally precedes the diagnosis of hidradenitis suppurativa. We present two new cases in which hidradenitis lesions developed before Crohn's disease, suggesting an overlap and supporting an association. Furthermore, in one of these cases, treatment with infliximab resulted in marked improvement in both the Crohn's disease and hidradenitis suppurativa.

KEY WORDS

hidradenitis suppurativa, Crohn's disease, inflammatory disease

Introduction

Crohn's disease (CD) and hidradenitis suppurativa (HS) are chronic, recurrent, inflammatory diseases of the epithelia. Onset usually occurs during young adulthood. In CD, the inflammation may involve any part of the gastrointestinal tract, and is characterized by discontinuous, transmural, inflammatory lesions of the gut wall (1). The characteristic clinical presentation of CD is abdominal pain and diarrhea, which may be complicated by intestinal fistulization and/or obstruction. HS is characterized by chronic, recurrent, inflammatory, painful, and suppurating lesions affecting hair follicles that are located on apocrine gland–bearing skin (e.g., the axillae and groin). These lesions often heal leaving scarring behind (2).

There are several similarities between CD and HS. The etiology of both diseases is complex and largely unknown, but it appears that the mechanisms behind these diseases are multifactorial, caused by an interaction of genetic and environmental factors, and characterized by a dysregulated local immune response (1, 2). Smoking has been identified as a risk factor for both diseases. Moreover, a clinical response to anti-TNF alpha therapy has been described for both diseases (1, 2).

An association between CD and HS has been suggested in previous reports (3–13; Table 1). We present two additional cases with comorbidity of these two diseases, which offer additional support for the hypothesis that these two diseases are associated.

Case report

Case 1

A 40-year-old female smoker presented with both HS and CD. Starting with the onset of a single abscess in an axilla when she was 26 years old, the patient subsequently experienced recurrent abscesses, nodules, and fistulas bilaterally in the axillae, groin, and submammary and anogenital regions. She was diag-

nosed with severe HS (Hurley stage 3, Sartorius score 99; Fig. 1). Unsuccessful treatments included multiple topical and systemic antibiotics as well as surgery. Two years after the onset of HS, she started experiencing daily, mucousy, non-bloody diarrhea and intermittent diffuse abdominal pain for a year before consulting a doctor. A colonoscopy was performed, which showed multiple ulcerations and the cobblestone pattern characteristic of CD. Long-term treatments included systemic prednisolone and azathioprine, which gave some relief but were not entirely effective. To date, anti-TNF-alpha therapy is the only treatment that has been able to control both this patient's CD and HS.

Case 2

A 44-year-old female smoker presented with both HS and CD. The patient was 38 years old when she first began experiencing recurrent nodules, abscesses, and fistulas, primarily in the genitofemoral region but also in the axillae and submammary area. She was clinically diagnosed with severe HS (Hurley stage 3, Sartorius score 66). Numerous attempts to treat the condition included the use of various types of topical and systemic antibiotics (tetracycline, clindamycin, rifampicin, and dapson), immunosuppressive drugs (cyclosporine, etanercept, infliximab, and methotrexate), topical resorcinol, and surgery. None of these treatments resulted in a satisfactory outcome.

At the age of 42, the patient presented with a 6-month history of intermittent diarrhea, vomiting, anorexia, weight loss, and fatigue. A sigmoidoscopy was performed, which showed diverticulitis. A biopsy indicated diverticulitis and revealed inflammation with numerous granulomas characteristic of CD. A laparoscopy revealed an inflamed sigmoid colon. An X-ray of the colon showed stricture of the sigmoid colon. The patient underwent surgery (sigmoidectomy) and subsequently experienced a number of postoperative complications, including fascial rupture and anastomosis leakage.

Discussion

This report describes the comorbidity of CD and HS in two patients, reinforcing the likelihood that there is an association between these two diseases. The shared characteristics in both cases reported were: 1) the patients were females, 2) HS was diagnosed before CD, and 3) the treatment of HS was difficult. From a clinical point of view, these observations show that physicians should be alert whenever HS patients display gastrointestinal symptoms. It is advisable to perform a colonoscopy to exclude CD in these patients.

Recently, nine case reports and two retrospective studies have suggested an association between HS and CD (3, 4, 6–12; Table 1). In contrast to our cases, most of the previously reported cases presented with the diagnosis of CD prior to that of HS, and a majority of these patients were men. In addition to case reports, two retrospective studies have been performed addressing this topic. In a pilot study, Van der Zee et al. (13) interviewed 102 CD patients about recurrent painful boils in the axillae and/or groin (Table 1). The

Table 1. Summary of reported patients with both Crohn's disease and hidradenitis suppurativa

Author	Year	Study design	Patients (*n*)	A: HS → CD B: CD → HS	Sex
C Gower -Rousseau	1992	Case report	3	B, B, B	M
MK Roy	1997	Case report	1	A	M
EV Tsianos	1995	Case report	1	B	M
NP Burrows	1992	Case report	2	A, A	M, F
LS Ostlere	1991	Case report	3	B, B, B	F, F, M
M Roussomoustakaki	2003	Case report	1	A	F
HH van der Zee	2009	Cross-sectional	102	*	M, F
F Martinez	2001	Case report	1	B	F
RL Attanoos	1993	Case report	3	B, A, B	F, M, M
AA Kafity	1993	Case report	1	B	M
JM Church	1993	Retrospective review	61	**	M, F

*Note. *The prevalence of HS was estimated in 102 CD patients. **The occurrence of CD was retrospectively reviewed in 61 HS patients. A: HS → CD means that the diagnosis of HS predated that of CD. B: CD → HS means the diagnosis of CD predated that of HS.*

study showed that 17% of CD patients had a history compatible with HS, again suggesting an association between HS and CD. Furthermore, Church et al. (5; Table 1) performed a retrospective review of hospital records for 61 HS patients and found that 24 also had a diagnosis of CD. The diagnosis of CD predated that of HS by an average of 3.5 years.

When HS affects the anogenital region, differentiation from CD can be difficult because both CD and HS can present as fistulas and sinuses, and histologically as granulomas (either as foreign body-type or epithelioid-type; 10). Therefore, the anogenital HS may only be distinguished from CD based on a history of recurrent painful boils in apocrine gland–bearing skin. In both of our cases, the diagnosis of HS is reliable, because clinically characteristic HS manifestations were observed in both the anogenital region and axillae.

The comorbidity or association of these two diseases may have an impact on treatment. It is of great importance for both the dermatologist and the gastroenterologist to be aware of this association, because a lack of awareness may lead to inappropriate or insufficient treatment of both diseases.

The TNF-alpha inhibitor infliximab has been shown to be effective in the treatment of both HS and CD (1, 8, 10). Infliximab is used when conventional medical and surgical treatments fail. In our first case, improvement in both HS and CD was observed when infliximab was administered. These observations reinforce the link between HS and CD and suggest a shared inflammatory pathway.

Although our findings and also previous reports suggest an association between HS and CD, more prospective studies are warranted in order to establish this association and its underlying pathogenesis.

REFERENCES

1. Jemec GB, Revuz J, Leyden J. Hidradenitis suppurativa, 1st ed. Heidelberg: Springer; 2006. 204 p.

2. Revuz J. Hidradenitis suppurativa. J Eur Acad Dermatol Venereol. 2009;23:985–98.

3. Attanoos RL, Appleton MA, Hughes LE, et al. Granulomatous hidradenitis suppurativa and cutaneous Crohn's disease. Histopathology. 1993;23:111–5.

4. Burrows NP, Jones RR. Crohn's disease in association with hidradenitis suppurativa. Br J Dermatol. 1992;126:523.

5. Church JM, Fazio VW, Lavery IC et al. The differential diagnosis and comorbidity of hidradenitis suppurativa and perianal Crohn's disease. Int J Colorectal Dis. 1993;8:117–9.

6. Gower-Rousseau C, Maunoury V, Colombel JF, et al. Hidradenitis suppurativa and Crohn's disease in two families: a significant association? Am J Gastroenterol. 1992;87:928.

7. Kafity AA, Pellegrini AE, Fromkes JJ. Metastatic Crohn's disease. A rare cutaneous manifestation. J Clin Gastroenterol. 1993;17:300–3.

8. Martinez F, Nos P, Benlloch S et al. Hidradenitis suppurativa and Crohn's disease: response to treatment with infliximab. Inflamm Bowel Dis. 2001;7:323–6.

9. Ostlere LS, Langtry JA, Mortimer PS et al. Hidradenitis suppurativa in Crohn's disease. Br J Dermatol. 1991;125:384–6.

10. Roussomoustakaki M, Dimoulios P, Chatzicostas C, et al. Hidradenitis suppurativa associated with Crohn's disease and spondyloarthropathy: response to anti-TNF therapy. J Gastroenterol. 2003;38:1000–4.

11. Roy MK, Appleton MA, Delicata RJ, et al. Probable association between hidradenitis suppurativa and Crohn's disease: significance of epithelioid granuloma. Br J Surg. 1997;84:375–6.

12. Tsianos EV, Dalekos GN, Tzermias C, et al. Hidradenitis suppurativa in Crohn's disease. A further support to this association. J Clin Gastroenterol. 1995;20:151–3.

13. Van der Zee HH, van der Woude CJ, Florencia EF, et al. Hidradenitis suppurativa and inflammatory bowel disease: are they associated? Results of a pilot study. Br J Dermatol. 2010;162:195–7.

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