Hidradenitis Suppurativa and Crohn’s Disease: Two Cases with Different Disease Courses

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ABSTRACT
Hidradenitis suppurativa is a rare chronic/recurrent, inflammatory and suppurative disease that affects apocrine gland-bearing areas of the skin, including the axillae, inframammary fold, inguinal and anogenital regions. Few reports have described the co-occurrence of Hidradenitis suppurativa and Crohn’s disease and the challenge in managing patients with both conditions. We describe two cases in which HS developed during the course of Crohn’s disease in one patient, while it preceded the onset of Crohn’s disease in the other case. Both patients showed poor response to biological therapy although one case initially responded well to induction therapy with infliximab.

Keywords
Adalimumab; Crohn’s disease; Hidradenitis suppurativa; Infliximab.

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INTRODUCTION

Hidradenitis suppurativa (HS) and Crohn’s disease (CD) are chronic, recurrent, inflammatory diseases that affect the epithelia. In CD, the inflammation may involve any portion of the gastrointestinal tract, and patients typically present with abdominal pain and diarrhoea, which may be complicated by intestinal fistulisation\[3\]. On the other hand, patients with HS typically present with chronic, recurrent, painful, inflamed, and suppurating lesions involving the hair follicles in apocrine gland-bearing areas of the skin\[2\].

Hidradenitis suppurativa can mimic cutaneous CD, which is both an associated disease and differential diagnosis of HS\[3\]. While HS and CD share many similarities, the aetiology of both diseases is complex and for the most part unknown. The pathophysiological mechanisms underlying both conditions are multifactorial and involve genetic and environmental factors, characterised by impaired local immune response\[1\]-\[4\]. Consequently, it is difficult to treat HS, and in most cases, response to treatment is either poor or partial, and the recurrence rate is high\[2\].

Multiple medical treatments for HS have been proposed. Clindamycin lotion or resorcinol cream have been shown to be effective in patients with mild disease. Tetracyclines are a first-line systemic option in patients with severe or extensive disease; a combination of clindamycin and rifampicin should be administered after first-line therapy. Nevertheless, the rate of recurrence is high after clindamycin and rifampicin combination therapy is discontinued\[3\]. While long-term therapy with acitretin (a retinoid compound) is feasible, it might have teratogenic effects when administered to women of reproductive age. Anti-inflammatory drugs, including cyclosporine, dapsone or fumarates, have been proposed for the treatment of HS. However, there is little evidence of their effectiveness. Biologics (such as adalimumab or infliximab) are the next option in cases where most common treatments have failed\[3\].

To date, no evidence-based effective treatment is available for HS, and therapeutic results are often disappointing because of the lack of targeted therapies. We present the cases of two patients with HS and CD who had different disease courses and showed varied clinical response to medical treatment.

CASE REPORTS

Case One

The patient is a 23-year-old Bangladeshi female diagnosed with ileal CD at the age of eight years. She initially presented with abdominal pain, diarrhoea and bloody stools. Endoscopic findings included a large ulcer and inflammation of the ileal mucosa, while a histopathology examination demonstrated edema, inflammation, architectural crypt changes, crypt abscesses, and transmural extension of the inflammation. Prior treatments for her CD included steroids, azathioprine, as maintenance therapy, and intermittent courses of prednisolone during relapse periods.

At the age of 18 years, she was started on intravenous infliximab 5 mg/kg for CD. Although the patient was compliant to treatment, she developed painful, suppurating skin lesions in both axillae, groin and chest. A dermatology consultation was requested and histopathological examination was offered. A skin biopsy specimen was obtained from her axillary region and groin. Histopathology typical of HS included dense lymphocytic infiltrates around the hair follicle and inflammatory cells around apocrine glands. The follicle contained pus. A chest X-ray and purified protein derivative tuberculin test were ordered to rule out tuberculosis.

Additional treatment, which comprised a combination of topical steroids and antibiotics (for 2 weeks) and oral prednisolone 15 mg daily, was applied. Subsequently, after nine cycles of intravenous infliximab (administered over a period of two years), adalimumab 40 mg biweekly was prescribed, as infliximab did not cause substantial resolution of her CD lesions. Infliximab and infliximab antibody levels were not measured.

Six months later, adalimumab was increased to 80 mg biweekly after the patient developed complicated fistulising perianal disease; the treatment was interrupted when her lesions did not improve. Surgical drainage with seton placement was offered. A repeat surgery was performed two months after the first procedure.

During the next four years, she underwent several procedures, including perianal drainage, surgical drainage of the left armpit and surgical excision of the lesions in the left axilla.

Case Two

The patient is a 24 year-old Saudi female diagnosed with HS at the age of 14. Histopathology findings of HS included infundibular hyperkeratosis and a dermis with an inflammatory cell infiltrate, granulation tissue and subcutaneous abscess. Her lesions involved the chest, inframammary region, groin, and thighs (Fig. 1 A and B). A chest X-ray and purified protein derivative tuberculin test were negative for tuberculosis. She underwent medical treatment comprising oral isotretinoin and alternating short courses of oral amoxicillin / clavulanic acid and metronidazole over a period of 18 months. Maintenance therapy included azathoprine and methotrexate.

At age 20, six years after HS diagnosis, a colonoscopic examination revealed irregular ulceration, numerous pseudopolyps, and loss of haustiations in the ileocolonic segment, suggestive of CD. A biopsy of the mucosa revealed cryptitis, loss of goblet cells, and inflammatory cells in the lamina propria. The patient was treated with steroids. At the age of 21, she was started on subcutaneous adalimumab 40 mg/kg for her HS lesions, but three years later, the treatment was interrupted due to poor clinical response despite good compliance. Drug level monitoring was not performed as it was unavailable. Intravenous infliximab 5 mg/kg was subsequently administered. The patient showed good response to initial induction therapy (Fig. 2), but her skin lesions got worse even after the dose of
FIGURE 1.
(A) Comedones and post-inflammatory hyperpigmentation due to recurrent abscesses at the left inframammary area; (B) Thigh with erythematous draining sinus before treatment.

FIGURE 2.
Left thigh with decreased erythema and no drainage after initial induction therapy with infliximab (5 mg/kg).
infliximab was increased to 10 mg/kg. Further treatment was continued abroad upon the patient’s request.

**DISCUSSION**

This report describes two cases of CD and HS. While our patients shared some similarities—same gender and poor response to biological therapy—one patient developed CD prior to HS diagnosis and the other developed CD after HS diagnosis. Associations between CD and HS have been suggested in other studies\(^6\), with one report\(^6\) stating that 17% of patients with CD had a history consistent with HS.

The first patient in our report had severe HS that was complicated by perianal fistulisation. In practice, cutaneous CD must be differentiated from perianal HS given that perianal lesions of CD may mimic HS: both diseases can clinically present as fistulas and histologically as granulomas\(^5\). An association has also been suggested between HS and CD based on immunological analysis of infiltrating T cells compared to peripheral blood\(^5\). In their report, Giudici et al.\(^6\) observed an accumulation of CD161+ T lymphocytes in CD fistulae and HS lesions. It is known that CD161+ T lymphocytes are rich in T helper 17 (Th17), Th17/ T helper 1 (Th1), and non-classic Th1 phenotypes\(^6,9\). Previous studies\(^6,9\) suggested that Th1 and Th17 were implicated in the pathogenesis of many inflammatory diseases. The authors also demonstrated that in the presence of interleukin 12, Th17 cells could switch to Th1 through a state in which the cells were able to produce both interleukin 17A and interferon y. The association of HD and CD might thus have an impact on treatment, underscoring why it is important for dermatologists and gastroenterologists to be aware of this association.

One report\(^9\) that supported the co-occurrence of HS and CD suggested that adalimumab, which is approved for CD treatment, might also be a safe and effective therapeutic option in the treatment of HS. Unfortunately, the first patient in our report did not show a dramatic response to adalimumab therapy, prompting us to believe that a better approach would have been to offer surgery after the failure of infliximab and conventional therapy (corticosteroids, supports the use of infliximab to prepare HS patients for topical and systemic antibiotics). Another report “curative” surgery\(^14\); nevertheless, there are concerns the approach would have been to offer surgery after the failure patient in our report did not show a dramatic response to this class of biologics in patients with poor or no clinical response. While the use of antibiotics in HS is debatable, its use may relieve symptoms in patients with extensive disease progression\(^13\).

**CONCLUSION**

Taken together, our report further suggests an association between HS and CD. However, prospective studies have to be conducted to establish the nature of this association and the potential mechanisms underlying this association. Meanwhile, physicians treating patients with a possible association of both conditions should consider current available treatment options while keeping in mind that although HS might be associated with CD, the mechanism of inflammatory cascade may not involve the tumour necrosis factor pathway. Furthermore, physicians treating patients in whom they suspect an association of HS and CD should be encouraged to share their experiences with different therapeutic options, as these can help investigators explore new effective treatment strategies.

**Informed Consent**

Informed consent was obtained from the patients before submission of this case report for publication.

**Conflict of Interest**

The authors have no conflict of interest.

**Disclosure**

None of the authors received any type of commercial support either in forms of compensation or financial for this study. They have no financial interest in any of the products or devices, or drugs mentioned in this article.

**Ethical Approval**

Obtained.

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حالتان نادرة تجمعان بين التهاب الغدد العرقية القيحي ومرض كرون

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المستخلص

التهاب الغدد العرقية القيحي هو مرض نادر مزمن، ينتج عنه التهاب متكرر في الغدد العرقية للجلد وخاصه منطقة الأبط، ومنطقة ما تحت الثديين والمناطق الشرجية التناسلية، وقد بُنيت بعض الدرواس احتمالية وجود علاقة بين مرضى كرون (التهاب مزمن في الأمعاء) والتهاب الغدد العرقية القيحي، ونتج عن ذلك ت喙بادات كبيرة في علاج هذين المرضى من المجتمعين، ونصف حالاتن مختلفتين تجمع بين المرضى في بداية ظهورهما واستجابهما للعلاج، ففي الحالة الأولى ظهر التهاب الغدد العرقية القيحي بعد ظهور مرض كرون، أما في الحالة الثانية فقد سبق التهاب الغدد العرقية القيحي مرض كرون وكانت استجابة كلا المرضى ضعيفة بالنسبة للعلاج البيلوجي، على الرغم من أن أحدهما استجابة لهذا العلاج في مرحلته الأولى فقط.