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Hidradenitis Suppurativa and Cutaneous Crohn's Disease Without Intestinal Inflammatory Bowel Disease

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OBJECTIVES

- To recognize that hidradenitis suppurativa (HS) and Crohn's disease (CD) are comorbid conditions and may occur together
- To appreciate when extraintestinal manifestations of CD may be present despite absence of intestinal disease
- To employ a low threshold for referral to gastroenterology for evaluation
- To emphasize that early detection, intervention, and close monitoring are critical for management of HS patients at potentially higher risk of CD development.

INTRODUCTION

- Hidradenitis suppurativa is a chronic, debilitating, inflammatory disease of the hair follicle characterized by recurrent, painful nodules and abscesses that can progress to form fistulae, draining sinus tracts, and scarring
- Crohn disease is a subtype of inflammatory bowel disease (IBD) in which discontinuous areas of transmural granulomatous inflammation can involve any portion of the gastrointestinal tract
- CD can be limited and cutaneous in extent, although most often CD will also involve the gastrointestinal tract.
- "Knife-cut" fissures, which resemble lacerations, are highly characteristic of CD

CASE PRESENTATION

HPI:

- 32 y/o AA woman presents for follow-up on skin biopsies performed during a hospitalization for newly diagnosed HS. ROS negative for intestinal symptoms.
 - Skin biopsy:** neutrophilic dermatosis and edema with mixed suppurative and granulomatous dermatitis along a hair shaft, compatible with HS
 - Colonoscopy and rectal biopsies:** normal ileum and colon but did reveal perianal abscesses; gastroenterology deemed that the patient had no evidence of CD or ulcerative colitis.

Physical exam:

- Tender, large, erythematous, indurated, ulcerated plaques with purulent drainage to her abdominal fold, inguinal folds, gluteal cleft and perianal area
- Multiple erythematous ulcers and inflammatory nodules to her bilateral buttocks (**Figure 1**)
- Deep "knife-cut" fissure of her right inguinal fold (**Figure 2**)
- Superficial ulcers to her bilateral medial thighs

Diagnosis:

- HS and cutaneous Crohn's without intestinal IBD

Patient course:

- Infliximab infusions titrated up to a dose of 5mg/kg every 4 weeks with significant improvement
- She continues to be followed by gastroenterology, although she exhibits no symptoms of intestinal IBD to date

DISCUSSION

- HS and CD share overlapping clinical and pathological features: abscesses and nodules with fistula formation as well as disease responsiveness to tumor necrosis factor inhibitors; smoking and obesity are risk factors.
- Although both can form granulomatous lesions, HS is marked by keratin plugging and follicular rupture.
- 40% of patients with CD develop extraintestinal manifestations (EIM) and most commonly presents in the skin.
- Up to 25% of EIM precede the diagnosis of intestinal CD; however, it is more common to exhibit EIM subsequent to intestinal CD.
- CD specific cutaneous manifestations are lesions that share the same histopathological features as the intestinal disease
- "Knife-cut" fissures are considered to be pathognomonic for cutaneous CD
- Because these cutaneous manifestations can clinically resemble other dermatologic diseases, particularly HS, histopathology and a complete GI evaluation is necessary
- Despite the lack of bowel-related symptoms in these patients, prompt referral to a gastroenterologist is warranted, as progression of cutaneous CD to involve the bowel may be forthcoming.
- Both adalimumab and infliximab have shown efficacy in the treatment of CD and HS which may be beneficial in patients with overlapping disease.

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FIGURE 1



Figure 1: multiple erythematous punched out ulcers to bilateral medial buttocks

FIGURE 2



Figure 2: "knife-cut" fissure of the right inguinal fold