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.

• HS and cutaneous Crohn’s without intestinal IBD.

• She continues to be followed by gastroenterology, although she exhibits no symptoms of intestinal IBD to date.

• 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.

FIGURE 1

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

REFERENCES


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.

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• 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.