

Introduction

Crohn's disease (CD) is a chronic inflammatory bowel disease with known cutaneous manifestations. While most lesions occur in areas contiguous with the GI tract, metastatic CD is a rare form marked by granulomatous skin lesions at distant, non-contiguous sites. We present two cases highlighting the variable clinical presentation of metastatic CD.

Case 1

Clinical presentation

- 14-year-old male presented with a 2-month history of a rash localized to the intergluteal cleft
- Mild burning pain with bowel movements, otherwise asymptomatic
- No rashes or plaques elsewhere on the body
- Minimal improvement with hydrocortisone suppository (prescribed by GI), topical hydrocortisone 2.5% cream, and ketoconazole 2% cream
- Exam notable for erythematous pebbly plaque involving the intergluteal cleft (Figure 1)

PMH: Crohn's disease, diagnosed 4 months prior to presentation

Medications: Adalimumab 40 mg SUBQ every other week

Results

Punch biopsy, left intergluteal cleft r/o psoriasis vs cutaneous Crohn's

- Focal parakeratosis, epidermal hyperplasia, and moderately dense superficial and deep lymphohistiocytic infiltrate with scattered multinucleated histiocytes
- Granulomatous inflammation compatible with cutaneous Crohn's disease

Outcomes

- Upon further work-up, adalimumab levels were subtherapeutic and dosing frequency was increased



Figure 1: Pebbly plaque on the intergluteal cleft

Case 2

Clinical presentation

- 31-year-old female presented with a 4-month history of painful, open wounds involving the lower abdomen and inguinal folds
- 2-week history of melanotic stools, diarrhea, and abdominal pain
- Denied GI symptoms at prior visits, no personal or family history of IBD
- Negative HSV and VZV cultures
- Some improvement with prednisone 80 mg daily but flared upon tapering
- Exam notable for knife-cut fissures on the lower abdomen along the right lateral cesarean section scar and inguinal folds; overlying erythematous patch in the infra-abdominal fold (Figure 2, 3)

PMH: Hidradenitis suppurativa, Hurley Stage 2-3 diagnosed 2016

PSH: Transverse cesarean section in 2023, uncomplicated

Medications: Adalimumab 40 mg SUBQ weekly, prednisone 80 mg QAM, fluconazole 200 mg weekly

Results

Punch biopsy, right infra-abdominal r/o cutaneous Crohn's

- Scattered multi-nucleated giant cells in the inflammatory infiltrate
- PAS stain negative for fungal organisms
- Histologic changes non-specific, but compatible with cutaneous Crohn's disease in the appropriate clinical context

Outcomes

- Ongoing work-up for intestinal Crohn's with GI



Figure 2: Knife-cut fissure on the right lateral C-section scar with overlying erythema



Figure 3: Knife-cut fissure on the right inguinal fold with HS scarring

Teaching Points

- Metastatic Crohn's disease is defined by granulomatous skin lesions at sites non-contiguous to the GI tract and does not correlate to the severity of underlying intestinal disease
- Cutaneous manifestations may precede GI involvement by months to years, and the absence of GI symptoms does not exclude cutaneous Crohn's disease
- Diagnosis is primarily clinical, but skin biopsy can be a helpful, though often non-specific, adjunct

Discussion

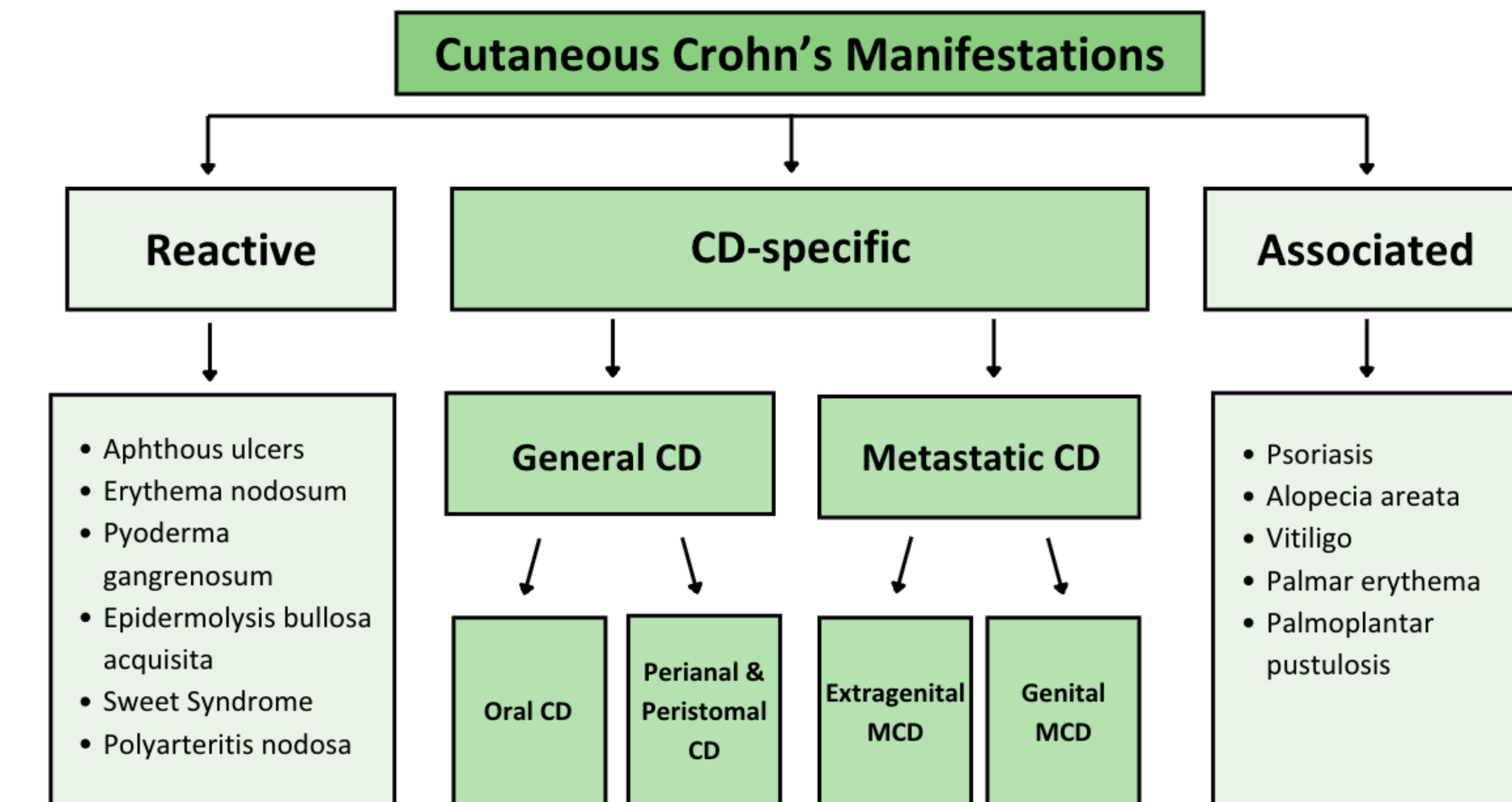


Figure 5: Classification of cutaneous Crohn's disease¹

Cutaneous CD manifestations include: (1) CD-specific lesions with granulomatous inflammation identical to intestinal disease, (2) reactive lesions that are immunologically mediated but lack granulomas, and (3) associated conditions linked by shared pathophysiology or epidemiology (Figure 5).^{1,2} General cutaneous CD occurs at sites contiguous with the GI tract, often presenting as lip swelling, oral ulcers, perianal skin tags, and fissures.^{1,2}

Metastatic CD (MCD) occurs at distant sites and may present with or without known GI disease. Genital MCD is most common, often presenting with isolated genital swelling.¹ Extragenital MCD is rare with heterogeneous clinical presentation, often with nodules or fissures on the legs, intertriginous areas, trunk, or abdomen.^{1,2} The "knife-cut" sign describes linear erosions seen in MCD, as seen in Case 2.¹

The diagnosis of MCD is clinical.³ Skin biopsy may support the diagnosis, but findings are often nonspecific. MCD is typically co-managed by dermatology and gastroenterology, and treatment may require adjustments to systemic Crohn's disease therapy. For cutaneous disease, first-line therapy includes topical, intralesional corticosteroids, or systemic corticosteroids.^{4,5} Second-line options include immunomodulators such as methotrexate or cyclosporine, or TNF-alpha inhibitors like infliximab or adalimumab.^{4,5}

References

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