Metastatic Crohn's disease: a cutaneous manifestation

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Introduction

Crohn's disease (CD) is a multisystem, granulomatous inflammatory disease that may involve any part of the gastrointestinal tract, from the oral cavity to the annus, and often presents with extra-intestinal involvement including the skin, eyes, joints, or hepatobiliary system. Cutaneous manifestations, being the most common extra-intestinal site, result from a variety of etiologies. In particular, characterization of lesions at distant sites, noncontiguous with bowel disease and with histopathological findings consistent with Crohn's disease, are referred to as metastatic Crohn's disease (MCD).

In this case, we present a patient with history of fistulizing disease, diagnosed with Crohn's disease in 2019 and receiving Upadacitinib therapy, who was found to have metastatic Crohn's disease.

Case report

A 40 year-old African-American male with a past medical history of ileocolonic and perianal CD, diagnosed in 2019 and receiving upadacitinib therapy, who presented due to history of fistulizing disease initially diagnosed as HS and characterized as constant, draining cysts localized to buttock. Previously, his disease course had been complicated by ileal perforation, psoas muscle abscess, and pelvic necrotizing fasciitis. At this time, the patient had been receiving upadacitinib for 6 months for CD although uncertain of any improvement since initiating treatment. Previous therapies were significant for ustekinumab, infliximab, and methotrexate all of which were discontinued due to the development of palmoplantar psoriasis.

On presentation, physical exam was significant for involuting nodules with large ostia and granulomatous appearing erythematous papules to the superior intergluteal cleft and scarring of several nodules to right lower buttock. Given his history and previous presentations, diagnosis of Hurley Stage III HS verses cutaneous CD was further investigated. At this time, a 4mm punch biopsy of the gluteal cleft was obtained and demonstrated granulomas composed of epithelioid histiocytes with admixed eosinophils predominantly located in the superficial papillary dermis, supporting the diagnosis of MCD. Two weeks later, physical exam revealed four granulomatous appearing nodules with scarring and drainage to right buttock. The patient was continued on upadacitinib and elected to proceed with two intralesional Kenalog injections to the right intergluteal cleft and initiate topical steroid solution. Repeat intralesional Kenalog injection was performed 6 weeks later, along with the initiation of steroid ointment in place of the solution.

Three months following his initial presentation, patient noted significant improvement in pain and spontaneous drainage of a previously painful nodule, with reported persistence of one chronically draining lesion. Physical exam was significant for two sinus tracts with active drainage in addition to scarred plaques with several sinus tracts to the right gluteal cleft. At this time, the patient is pleased with his improvement while receiving upadacitinib and is only requiring topical steroid therapy as needed.

Discussion

Although an exact pathogenesis of MCD remains undetermined, multifactorial mechanisms have been hypothesized, suggesting the influence of an antigen-driven granulomatous response at the level of the dermis, cross-reactivity between gut and skin antigens, and T-cell-mediated hypersensitivity reactions to unknown antigens localized in the skin. Currently, there are no definite guidelines for treatment, although partial improvement has been reported in regimens targeting the inflammatory cascade and immune suppression. Given its variability in presentation and inconclusive therapeutic management, MCD remains a challenging diagnosis, emphasizing the importance of increasing awareness and education across disciplines to improve recognition of the disease and, thus, minimize delays in treatment and reduce the risk of morbidity.