55-year-old man with ulcers in inguinal fold and intergluteal cleft found to have systemic Langerhans cell histiocytosis

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INTRODUCTION

This article describes the case of a patient with cutaneous ulcers who was found to have systemic Langerhans cell histiocytosis (LCH). This article includes the clinical, histology, and electron microscopy images, in addition to a description of the presentation, workup, and management of this rare disease in this patients, with a review of the literature.

CASE

A 55-year-old man presented to the Veterans Affairs New York Harbor Hospital for evaluation of ulcers in his left inguinal and intergluteal folds that had been progressing over several months (Fig 1). During a screening colonoscopy for workup of worsening anemia 1 month prior, a polyp was excised and pathology revealed an unusual histiocytic infiltration seen on pathology. Positron emission tomography–computed tomography revealed hypermetabolic cutaneous foci involving the left inguinal and perianal regions corresponding to sites of cutaneous ulceration. The patient described the ulcers as asymptomatic and gradually enlarging in size. He did not have drainage or pain from the affected areas. He also had no sexual activity, nausea, vomiting, abdominal pain, blood in stool, fevers, weight loss, or other constitutional symptoms.

Examination of the patient's left inguinal fold and intergluteal cleft revealed well-demarcated ulcers with rolled cobblestoned borders and fibrinous granulation tissue at the ulcer base. The lesions were nontender on palpation without an associated odor, erythema, or vesicles. There was shotty inguinal lymphadenopathy. On the soft oral palate, exophytic verrucous plaques were noted.

Abbreviation used:

LCH: Langerhans cell histiocytosis

A complete blood count with differential analysis, basic metabolic panel, and rapid plasma reagin were unremarkable. On histopathologic examination, both skin and colon biopsies showed a dense proliferation of histiocytes and lymphocytes. Immunostaining was positive for S100, CD1a, and CD4 and weakly positive for CD45 and CD68 (Fig 2). CD20 and CD3 were negative. Electron microscopy of the skin biopsy showed Birbeck granules within the cytoplasm of a Langerhans cell (Fig 3). The skin lesion was negative for *BRAF* V600 mutations, including V600E, V600K, V600D, V600R, V600A, V600G, and V600M. A bone marrow biopsy was negative for CD1a and S100.

DISCUSSION

The diagnosis was the systemic LCH in an adult patient. LCH represents a spectrum of diseases characterized by clonal proliferations of Langerhans cells.[1]

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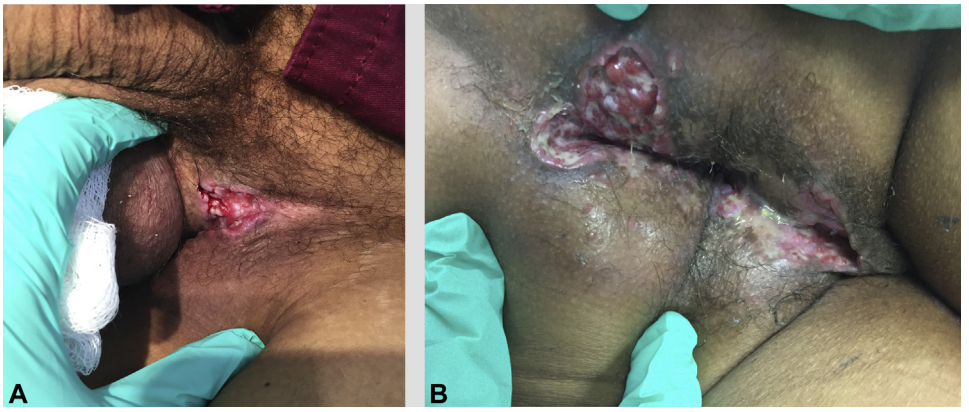

Fig 1. A and **B**, Well-demarcated inguinal and intergluteal ulcers found during physicial examination were nontender on palpation.

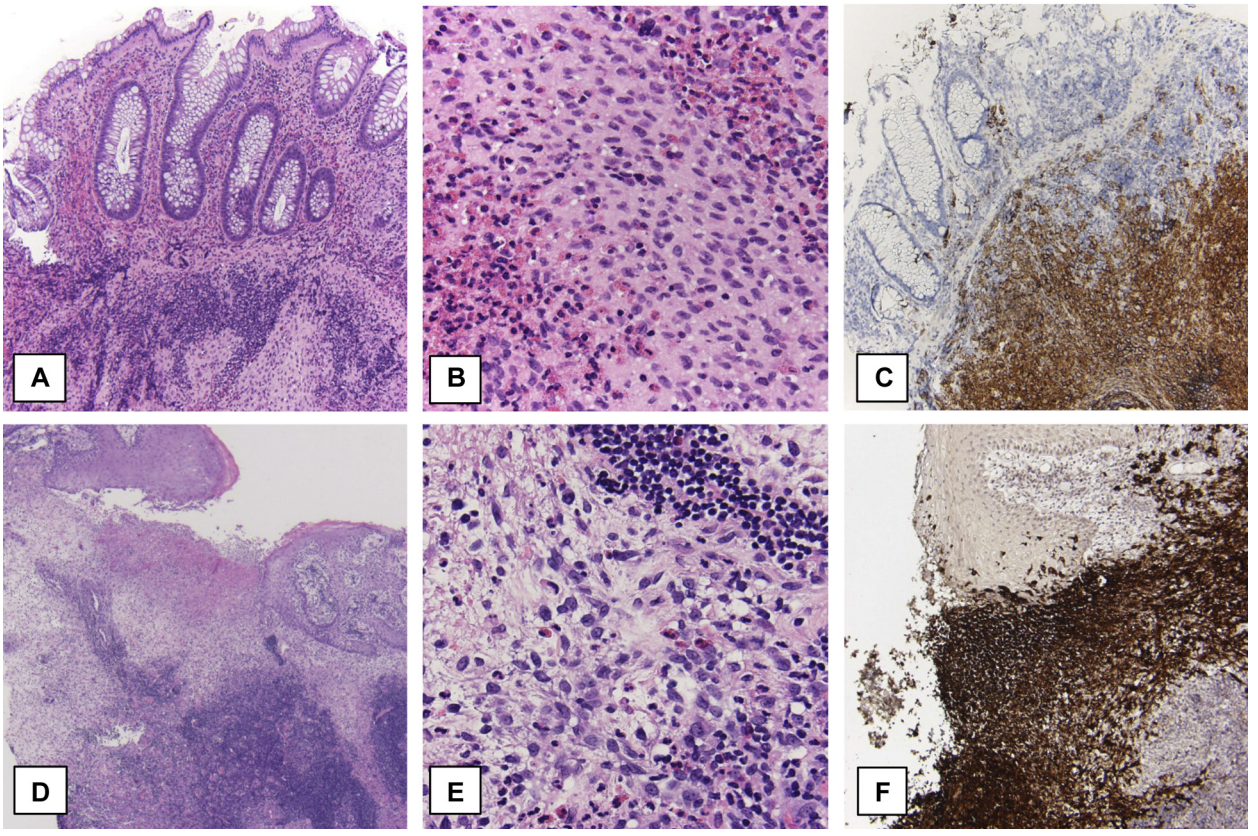

Fig 2. Colon and skin histopathology of patient with Langerhans cell histiocytosis. **A**, Polypoid colonic lesion shows a dense proliferation of histiocytes in the submucosa with eosinophilic abscesses in addition to neutrophilic and lymphocytic infiltrates. **B**, Oval histiocytes approximately 15 μm characterized by folded, indented or lobulated nuclei with fine chromatin, inconspicuous nucleoli, and thin nuclear membranes. The cytoplasms are moderately abundant and slightly eosinophilic. **C**, Langerhans cell histiocytosis involvement at colon. **D**, Skin biopsy reveals similar infiltrate to that of the colon specimen. **E**, Histiocytes with large, pale, folded or lobulated (often reniform), vesicular nuclei and abundant, slightly eosinophilic or amphophilic cytoplasms. **F**, Histiocytic infiltrate. (**A**, **B**, **D**, and **E**; Hematoxylin-eosin stain; **C** and **F**; CD1a stain; original magnification: **A**, **C**, **D**, and **F**, X2; **B** and **E**, X20.)