

Ulcerative cutaneous plasmacytosis



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INTRODUCTION

Cutaneous plasmacytosis is a rare entity of unknown etiology characterized by the benign proliferation of polyclonal plasma cells.^{1,2} Classically, it presents as reddish-brown plaques and papulonodules favoring the trunk and face. The disorder has been predominantly identified in people of Asian descent, although scattered cases involving patients of other ethnicities have been reported.³ As yet, no standardized therapy has been established, and the disease typically follows a chronic and benign clinical course. Here we describe a unique case of treatment-responsive cutaneous plasmacytosis presenting as ulcerative nodules in a Central American patient.

CASE REPORT

A 77-year-old Honduran woman presented with a 2-week history of new-onset, asymptomatic ulcers on her abdomen. Lesions began as small pustules and evolved into ulcers. She was otherwise well and denied any fevers, chills, malaise, or weight loss. The patient had no history of trauma, immunobullous disease, or autoimmune disorder. Her medical history included diabetes and hypertension. Travel history was significant for a recent month-long stay in Honduras. Medications included metformin, sitagliptin, amlodipine, metoprolol, and aspirin. At the time of examination, she was afebrile, with normal blood pressure and heart rate. Skin examination was notable for several well-circumscribed nontender ulcers of varying sizes, each with a bright red base of granulation tissue (Fig 1). There was no palpable lymphadenopathy.

Biopsy of an ulceration found a moderately dense superficial and deep perivascular and

Abbreviation used:

IL: interleukin

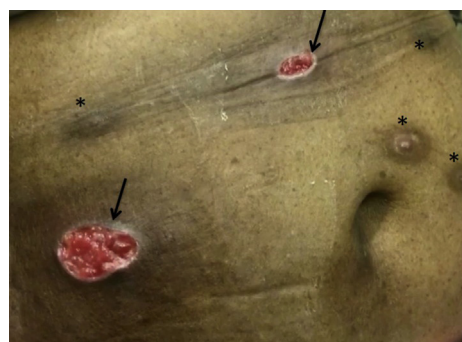


Fig 1. Well-circumscribed nontender large and small ulcers with bright red base of granulation tissue (arrows). Hyperpigmented plaques denoting healed ulcerations (asterisks).

interstitial infiltrate comprised predominantly of plasma cells lacking cytological atypia (Fig 2). Immuno-histochemical analysis found a normal κ/λ light chain ratio, and no microorganisms were identified by Gram, Periodic acid–Schiff, acid-fast bacillus, or *Treponema pallidum* stains. Molecular studies failed to reveal a clonal immunoglobulin gene rearrangement.

Laboratory workup including serum and urine protein electrophoresis, complete blood count, chemistry panel, liver function, lactate dehydrogenase, HIV, abdominal ultrasound scan, and dual-energy x-ray absorptiometry scan were unremarkable, supporting a diagnosis of cutaneous plasmacytosis without systemic involvement.

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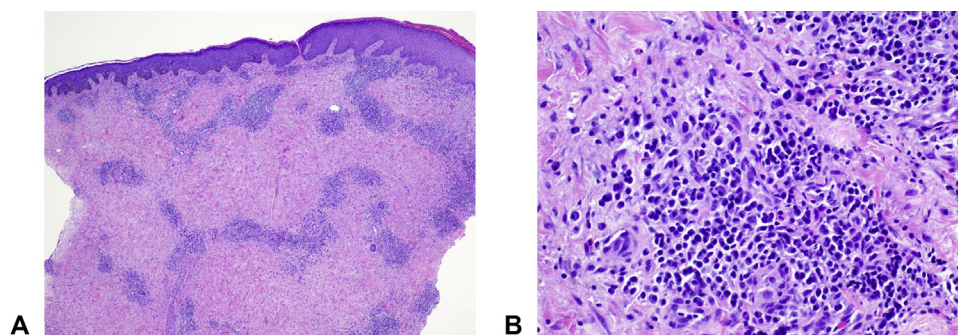


Fig 2. Histopathologic examination shows slight epidermal acanthosis, mild spongiosis, orthokeratosis and hypergranulosis overlying a moderately dense superficial and deep perivascular and slightly interstitial infiltrate comprised predominantly of plasma cells with admixed lymphocytes and a few histiocytes. Most plasma cells have uniform nuclei without cytological atypia. (Hematoxylin-eosin stain; original magnifications: **A**, $\times 4$; **B**, $\times 20$.)

Table I. Cases of cutaneous plasmacytosis in which ulceration was reported

Study	Age	Sex	Clinical presentation	No. of lesions	Location	Treatment
António et al ⁴	67	F	Ulcerated erythematous plaque	1	Face	Hydrocortisone cream every night at bedtime for 1 mo
Cerottini et al ⁵	90	M	Ulcers with diffuse, patchy hyperpigmentation	Multiple	Legs	—
Khullar et al ⁶	45	M	Hyperpigmented plaque with ulcerated nodule	1	Upper back	Clobetasol propionate 0.05% cream twice a day for 4 weeks + excision

The ulcers were treated with monthly intralesional injections of 10 mg/mL of triamcinolone and twice-daily application of topical mupirocin ointment. Upon healing, maintenance therapy with fluocinonide daily was initiated to the affected areas.

DISCUSSION

Here we describe a patient that presented with ulcerations in lieu of the papulonodules and plaques characteristic of cutaneous plasmacytosis. A report by António et al⁴ identified a patient who similarly presented with ulceration as the sole manifestation of cutaneous plasmacytosis. As with our patient, there were no systemic symptoms of disease, and the ulceration responded to treatment with subsequent healing. This ulcerative presentation and responsiveness to treatment deviates from the classic depiction of cutaneous plasmacytosis in Asian patients. It is plausible that such cases represent a unique variant of the disease distinct from the Asian variant. At this time, we have identified only 3 cases of cutaneous plasmacytosis in which ulceration was reported (Table I).⁴⁻⁶ Additional patients are needed to further investigate this hypothesis.

Cutaneous plasmacytosis typically follows a chronic, benign course, but a few cases have been reported to progress to systemic plasmacytosis. The most common extracutaneous manifestations are lymphadenopathy and polyclonal hypergammaglobulinemia.⁷ Renal amyloidosis, interstitial pneumonia, and hepatosplenomegaly occur less frequently.^{7,8} Serum immunoglobulin levels greater than 5000 mg/dL and bone marrow plasma cell percentage greater than 7% portend a more aggressive clinical course.³

Although the etiology of cutaneous plasmacytosis remains unknown, it has been postulated that the cytokine interleukin (IL)-6 is involved in pathogenesis. IL-6 promotes antibody production and drives the differentiation of mature B cells into plasma cells. Importantly, overexpression of IL-6 in mice results in a plasmacytosis.⁹ Accordingly, serum elevations of IL-6 have been reported in patients with cutaneous and systemic plasmacytosis.¹⁰ It is plausible that some type of insult, such as infection, autoimmune dysregulation, or genetic mutation, induces a localized or systemic increase in IL-6 that results in plasmacytosis.

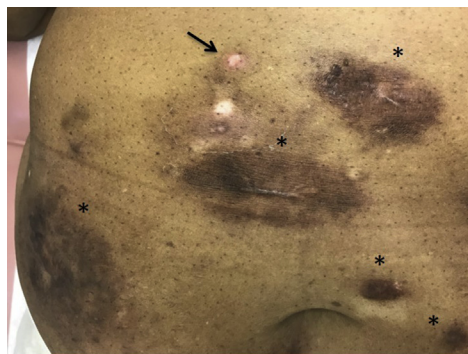


Fig 3. Resolution of ulcers with postinflammatory hyperpigmentation (asterisks). Four to six weeks after intralesional triamcinolone injection. Small erosion from a resolving ulcer (arrow).

Although there is no standardized treatment, a trial of topical and systemic corticosteroids, tacrolimus, psoralen ultraviolet A, photodynamic therapy, and pulsed dye laser are reported with varying success.¹¹⁻¹³ Our patient responded well to a combination of intralesional triamcinolone and mupirocin with complete healing of the lesions within a few months (Fig 3). Fluocinonide cream was then used as needed for maintenance therapy and to ameliorate pruritus associated with the healing ulcers. At the time of this report, one and a half years after presentation, the patient has had no associated systemic signs or symptoms.

Ulcerative cutaneous plasmacytosis is a unique manifestation of an already extremely rare disease. The identification of additional cases with this presentation will enable the further characterization of this reactive process and its clinical course.

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