



Cutaneous plasmacytosis resembling pityriasis rosea in a 66-year-old white woman: A rare disease presenting in an unusual patient demographic

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INTRODUCTION

Primary cutaneous plasmacytosis is a disorder of unknown etiology characterized by reddish brown papules and plaques primarily seen on the trunk and most commonly seen in middle-age men of Japanese descent. The disorder was first described by Yashiro¹ in 1976 and later termed *cutaneous plasmacytosis* by Kitamura et al² in 1980. Approximately 70 cases of cutaneous plasmacytosis have been reported in the literature, almost exclusively in the Asian population. Cutaneous plasmacytosis follows a chronic and mostly indolent course with pathologic features of dermal infiltrates with mature polyclonal plasma cells. Rare cases of systemic plasmacytosis with findings of lymphadenopathy, organomegaly, polyclonal hypergammaglobulinemia, interstitial pneumonia, and interstitial nephritis have been reported.³ Close monitoring with serial surveillance is essential for patients with cutaneous plasmacytosis. We describe a case of primary cutaneous plasmacytosis occurring in an unusual demographic, an older white woman.

CASE REPORT

A 66-year-old white woman of Polish descent presented with a 2-year history of asymptomatic red-to-brown, scattered, nonscaly, oval thin plaques that followed skin tension lines similar to pityriasis

Abbreviations used:

HHV-8: human herpes virus 8
IL: interleukin

rosacea. These lesions involved the upper back, chest, abdomen, neck, and axilla but spared the face and lower extremities (Figs 1 and 2). The patient was otherwise healthy and denied fever, malaise, night sweats, weight loss, or other constitutional symptoms. No hepatosplenomegaly or lymphadenopathy was appreciated.

Histopathologic examination found brisk papillary and mid-dermal perivascular and periadnexal inflammatory infiltrate of plasma cells and occasional lymphocytes (Fig 3). Kappa and λ in situ hybridization found a polyclonal proliferation. Findings from immunohistochemical studies and in situ hybridization were negative for spirochetes, human herpes virus 8 (HHV-8), and Epstein-Barr–encoded RNA.

Complete blood count with differential, complete metabolic panel, antinuclear antibody, anti-SSA/SSB antibodies, fluorescent treponemal antibody absorption test, serum and urine protein electrophoresis, β -2 microglobulin, and interleukin (IL)-6 levels were all within normal limits. Flow cytometry found normal serum levels of IgG, IgA, and IgM. Subclasses

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of IgG1, IgG2, IgG3, and IgG4 were all within normal limits. The κ free light chain level was elevated at 37.5 mg/L (ref. 3.3–19.6); however, the κ/λ free light chain ratio remained within normal limits at 1.54, and serum and urine immunofixation studies found no monoclonal immunoglobulins. Computed tomography of the chest, abdomen, and pelvis did not find any masses, lymphadenopathy, or lytic bone lesions. Based on the above findings, primary cutaneous plasmacytosis was diagnosed. The patient declined further treatment at this time because of the asymptomatic nature of the cutaneous plasmacytosis.

DISCUSSION

This case highlights the unusual presentation of cutaneous plasmacytosis in a white woman. Approximately 10 cutaneous plasmacytosis cases worldwide have been described in the literature occurring in the white population, with more than half of those cases coming from European countries such as Spain, Italy, and Germany. To our knowledge, our case represents only the fourth documented occurrence of cutaneous plasmacytosis in a white American. A review of 41 cases of cutaneous plasmacytosis in the Japanese population reported the male/female incidence ratio to be 1:0.6 and the age of onset to be 20 to 62 years; however, 5 cases of pediatric cutaneous plasmacytosis are reported in the literature in patients as young as 3 years.^{4,5}

Cutaneous plasmacytosis classically presents with multiple reddish brown ovoid-shaped papules and plaques found on the trunk. These lesions composed of mature polyclonal plasma cells are commonly distributed in a “Christmas tree–like” pattern. This truncal distribution and lesional shape can make cutaneous plasmacytosis difficult to distinguish from an atypical hyperpigmented variant of pityriasis rosea. Patients with cutaneous and systemic plasmacytosis often have constitutional symptoms including fatigue, weight loss, and fever.⁶ Similarly, a recent case series found prodromal symptoms, including fever and lymphadenopathy, are present in 59.6% of patients with pityriasis rosea.⁷ Pruritus is more common in pityriasis rosea but can occur in cutaneous plasmacytosis.^{8,9}

Systemic involvement of plasmacytosis can occur with plasmacytic infiltration of the skin and lymph nodes accompanied by polyclonal hypergammaglobulinemia, termed *cutaneous and systemic plasmacytosis*. Superficial lymphadenopathy was found in 58% and polyclonal hypergammaglobulinemia was found in 93%.⁴ Other less frequently involved sites include bone marrow, lung, liver, spleen, and kidney. In children, cutaneous plasmacytosis has been described as a separate entity called *isolated benign*



Fig 1. A 66-year-old white woman with multiple asymptomatic, reddish brown, ovoid thin plaques following skin tension lines on the back resembling pityriasis rosea.



Fig 2. Three nonconfluent linear-to-ovoid, reddish brown, thin plaques without scale in the right axilla.

cutaneous plasmacytosis, characterized by single skin lesions that show mature polyclonal plasmacytic infiltrate without the systemic findings of hypergammaglobulinemia and lymphadenopathy.⁷

Histopathology characteristically shows a dense superficial and deep perivascular and periadnexal dermal infiltrate of mature polyclonal plasma cells without atypia with few lymphocytes and histiocytes.⁸ Perineural infiltrates and lymphoid follicles with reactive germinal centers are less commonly reported.⁹ Immunohistochemistry shows polyclonal plasma cells with the existence of both κ and γ chain positivity.

The etiology of cutaneous plasmacytosis is unknown. One theory considers this disorder a variant of the reactive plasmacytic disorders; however, other

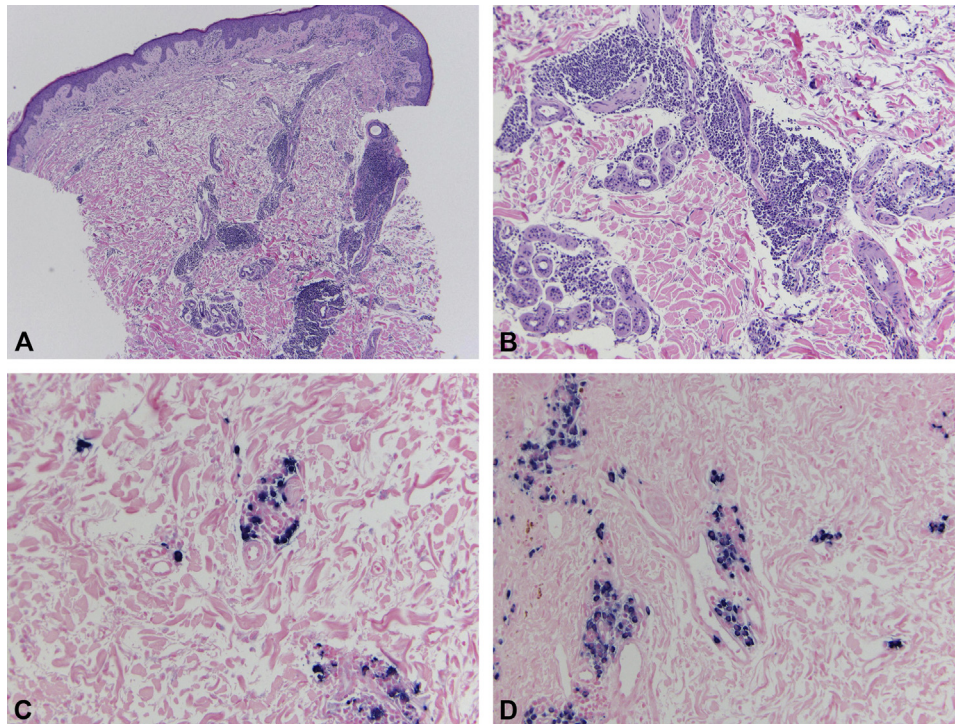


Fig 3. **A**, Skin biopsy section shows brisk superficial, deep, and periadnexal inflammatory infiltrate of plasma cells and occasional lymphocytes. **B**, Plasma cells. **C**, λ in situ hybridization. **D**, κ in situ hybridization. Cells stained positive for κ light chains and λ light chains. (Hematoxylin-eosin stain; original magnifications: **A**, $\times 4$; **B**, $\times 10$; **C** and **D**, $\times 20$.)

theories speculate a role in infectious, environmental, or genetic etiologies.^{4,10} Elevated levels of IL-6, a cytokine that plays a role in the differentiation of B cells to plasma cells, have been reported in some patients with cutaneous plasmacytosis. Some consider cutaneous plasmacytosis a variant of multicentric Castleman disease, as follicular hyperplasia and polyclonal plasma cell infiltrate with elevated levels of IL-6 may be seen in multicentric Castleman disease. Overproduction of IL-6 was attributed to cells that were latently infected with HHV-8. Because of the similarities, it was hypothesized that HHV-8 may also play a role in cutaneous plasmacytosis; however, most patients, including ours with histopathologically proven cutaneous plasmacytosis, are HHV-8 negative.¹¹

Cutaneous plasmacytosis is a chronic disorder with rare spontaneous regression and overall favorable prognosis. Multiple extracutaneous involvements, levels of hypergammaglobulinemia greater than 5,000 mg/dL, and concentration of plasma cells in the bone marrow greater than 7% are correlated with a more severe disease course. A few cases with fatal outcomes are reported in the literature with causes of death ranging from renal failure and leukemia to lymphoid interstitial pneumonia.⁴

Multiple treatment modalities have been attempted for cutaneous and systemic plasmacytosis with no standard treatment for this rare disease. Treatment options with variable success include antibiotics, topical and systemic corticosteroids, topical tacrolimus, chemotherapy, and anti-CD20 therapy.¹² Recent reports of low-dose thalidomide, narrow-band ultraviolet B therapy, mask-bath psoralen combined with ultraviolet A, and intralesional corticosteroid injections have shown improvement in cutaneous plasmacytosis.¹³⁻¹⁶

Although primary cutaneous plasmacytosis is a rare disease described in patients mainly of Japanese origin, it should remain on the differential diagnosis for patients of all races and sexes who present with the typical clinicopathologic features. Biopsy is necessary to distinguish primary cutaneous plasmacytosis from other dermatoses with similar presentations, including pityriasis rosea. Close monitoring and appropriate workup is essential to exclude systemic involvement.

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