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CASE REPORT

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Primary cutaneous plasmacytosis successfully treated with topical corticosteroids and psoralen plus ultraviolet A: A case report

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Key clinical message

We present a case of primary cutaneous plasmacytosis without systemic involvement. The patient received topical corticosteroids and psoralen plus ultraviolet A therapy, showing significant improvement. Continuous monitoring is underway despite the rarity of systemic manifestations.

KEYWORDS

cutaneous plasmacytosis, interleukin 6, phototherapy, plasma cell

1 | INTRODUCTION

Primary cutaneous plasmacytosis (PCP) is an uncommon cutaneous disorder characterized by the infiltration of plasma cells in the skin, representing a reactive process. The etiology of PCP is still unknown. It typically manifests as multiple infiltrative reddish-brown nodules and plaques, primarily affecting the trunk in adults. PCP has predominantly been reported in individuals of Japanese lineage, with fewer cases documented among Caucasians, Chinese, and Indians. This report presents a case of PCP in a 50-year-old woman who presented with isolated skin lesions on the trunk and upper back.

2 | CASE HISTORY/EXAMINATION

A 50-year-old woman presents with a progressively increasing rash on her body over the course of a year. She had no rash on her face and other sun-exposed areas. Physical examination revealed multiple discrete reddish-brown macules and papules on the chest wall and upper

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back (Figure 1A). Additional symptoms, including telangiectasis and a positive Darier's sign, were also noted. The patient did not report any other abnormal symptoms, such as dyspnea, arthralgia, nausea, vomiting, or diarrhea.

3 | METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS AND TREATMENT)

Regarding laboratory investigations, her complete blood count was otherwise normal, with no signs of atypical lymphocytes. Her kidney function and electrolyte levels were all within the normal limit. Serum protein levels showed an elevated globulin protein level of 3.7 g/dL (reference range: 1.3–3 g/dL). To further investigate the increase in globulin protein, a serum protein electrophoresis test was carried out. The test result indicated an abnormally high level of gamma globulin at 1.83 g/dL (reference range: 0.70–1.50 g/dL) without monoclonal gammopathy. Her IgG-4 levels were 148 mg/dL (reference range: 11–330 mg/dL).

Serology tests were negative for syphilis (VDRL and TPHA), human immunodeficiency virus (HIV), hepatitis B virus, and hepatitis C virus. The patient stated that she currently resides in Thailand and has never previously traveled abroad. Since Lyme disease is not endemic in Thailand, no serology tests for Borrelia were performed. The antinuclear antibody result showed negative.

A bone survey showed no bone lytic lesions. Bone marrow aspiration revealed a normal level of plasma cells. Further karyotype analysis was done on the bone marrow specimen, which showed an apparently normal female pattern [46, XX]. The red flag signs for multiple myeloma, including calcium elevation, renal failure, anemia, and bone lytic lesions, were all negative. The lymph node was not palpable. Computed tomography of the chest and abdomen revealed no evidence of aberrant lymphadenopathy or hepatosplenomegaly.

A skin biopsy taken from a reddish-brown macule on the left chest showed a piece of epidermis and dermis exhibiting superficial perivascular and periadnexal plasma cell infiltration alongside a small lymphocyte infiltrate (Figure 2). Immunohistochemical studies were used to suggest the polyclonality of the plasma cell infiltrates. CD38 and CD138 were utilized to confirm the presence of plasma cells (Figure 3).

4 | CONCLUSION AND RESULTS (OUTCOME AND FOLLOW-UP)

Based on the patient's history, physical examination, and the investigations mentioned above, the final diagnosis for the disease is primary cutaneous plasmacytosis. Interleukin 6 (IL-6) levels in the patient's blood were subsequently measured and found to be less than 1.5 pg/mL (reference range: 0–7) to explore its correlation to cutaneous plasmacytosis. She was treated with a combination of betamethasone dipropionate and psoralen plus ultraviolet A (PUVA), which showed a gradual decrease in the size and number of lesions after 17 sessions (Figure 1B). The PUVA regimen involved the administration of oral methoxsalen at a dosage of 0.6 mg/kg/day, taken 2 h prior to UVA exposure (Daavlin phototherapy, Bryan, OH, USA).

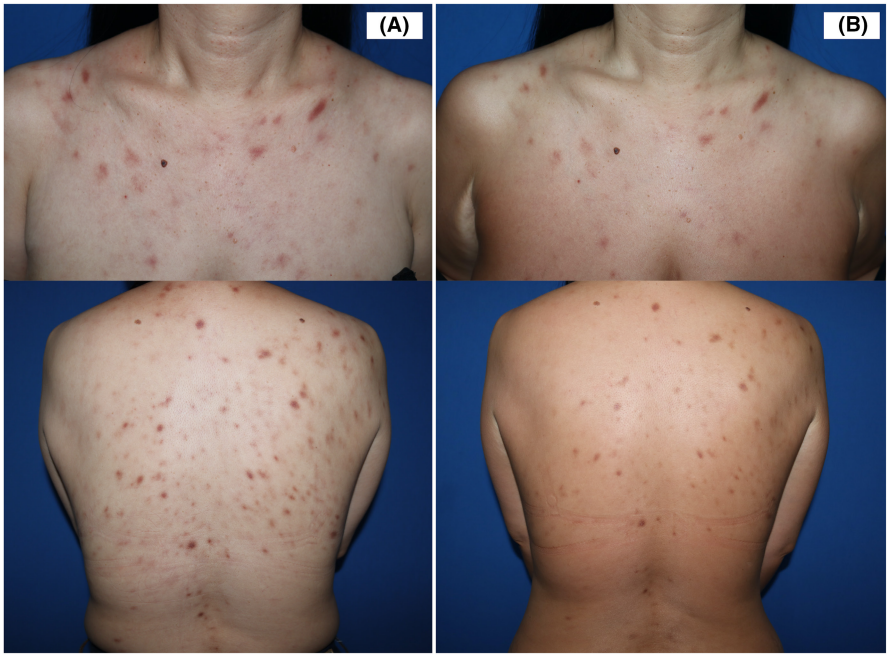

FIGURE 1 A 50-year-old woman experiencing (A) multiple reddish-brown macules and papules distributed on the chest wall and upper back (B) resolution after 17 sessions of topical corticosteroids and psoralen plus ultraviolet A.

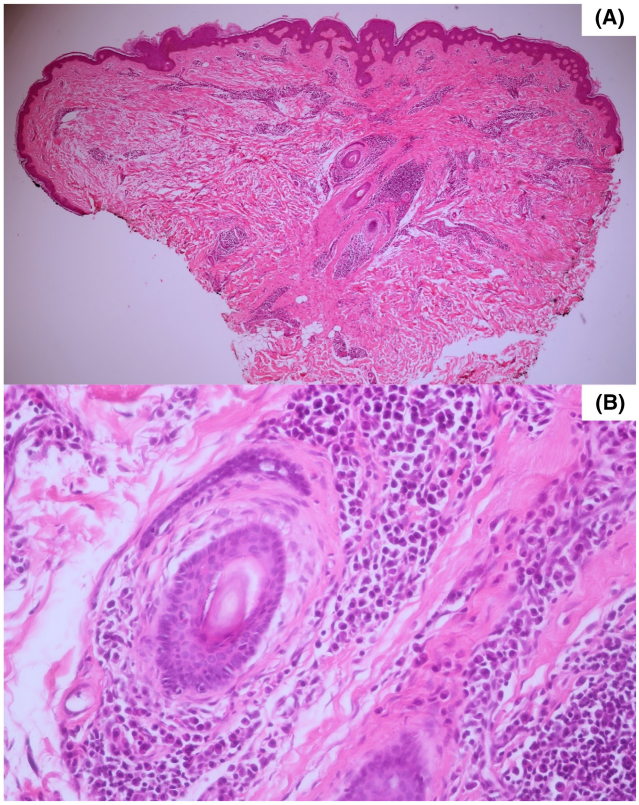

FIGURE 2 Hematoxylin and eosin stain showed a piece of epidermis and dermis exhibiting superficial perivascular and periadnexal plasma cells infiltration alongside small lymphocyte infiltrate (×40 [A] and ×400 [B]).

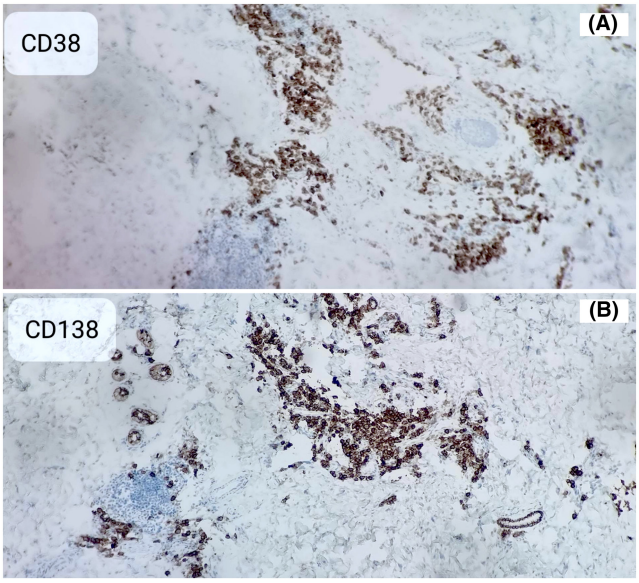

FIGURE 3 CD38 and CD138 were utilized to confirm the presence of plasma cells.

The whole-body treatment occurred twice per week, starting with a dosage of 1 j/cm^2 and incrementally increasing by 0.5 j/cm^2 until reaching 5 j/cm^2.

5 | DISCUSSION

Primary cutaneous plasmacytosis (PCP) is a rare reactive process cutaneous disorder. PCP is most commonly found in adults, with a slight male predominance, and is mainly described in patients of Japanese lineage. There have been fewer reports on Caucasians, Chinese, and Indians.[1] Multiple reddish-brown infiltrative macules, nodules, and plaques, often arranged in a linear or reticulate pattern, are the distinctive clinical characteristics of PCP.[2] The lesions are usually asymptomatic but may be pruritic in some cases. They commonly involve the trunk and extremities, and less frequently, the face and scalp.[3] Systemic involvement is rare, but extracutaneous involvement in cutaneous plasmacytosis has been documented, including hypergammaglobulinemia (up to 93%), lymphadenopathy (up to 58%), and involvement of the liver, spleen, lungs, and kidneys.[4–6]

The exact etiology of cutaneous plasmacytosis remains unknown. However, some studies suggest that it may be related to an abnormal immune response or an antigenic stimulus triggering clonal proliferation of plasma cells in the skin.[7] In addition, dysregulation of IL-6 production, which affects B-cell proliferation and maturation to plasma cells, is suspected to play a major role.[8] The association with various infections, such as human herpesvirus-8, Epstein–Barr virus, and hepatitis C virus, has been reported in a subset of patients.[9] However, the causal relationship between these infections and cutaneous plasmacytosis remains unclear.

The hallmark characteristic in histopathological changes of cutaneous plasmacytosis is the dense superficial and deep perivascular and periadnexal infiltration of mature polyclonal plasma cells within the dermis, often extending into the subcutaneous tissue, in the absence of a secondary cause of plasma cell infiltration.[10] The plasma cells typically exhibit eccentric nuclei with abundant basophilic cytoplasm, resembling the appearance of neoplastic plasma cells.[11] Immunohistochemical staining shows positive staining for CD138 and kappa or lambda light chains, confirming the plasma cell origin.[12] The presence of Russell bodies, which are eosinophilic intracytoplasmic inclusions, is a characteristic feature but not always present.[13] The isolated cutaneous involvement, however, is benign and persistent without spontaneous remission. However, there have been a few reports of PCP turning into lymphoma, but the incidence of malignancy transformation has not been established.[8] Additionally, cutaneous plasmacytosis may antedate the onset of idiopathic multicentric Castleman's disease (iMCD). In cases where a patient is diagnosed with cutaneous plasmacytosis lacking systemic symptoms, physicians should closely monitor and follow-up, recognizing the likelihood of iMCD progression.[14]

Asymptomatic cases of cutaneous plasmacytosis may not require specific treatment, and spontaneous resolution can occur in some patients. However, symptomatic patients or those with extensive involvement may benefit from therapeutic interventions. Various treatments, including topical and systemic medications, phototherapy, and radiotherapy, have all been explored, with varying degrees of effectiveness. Topical corticosteroids, topical 0.1% tacrolimus, systemic corticosteroids, immunomodulatory agents such as hydroxychloroquine, thalidomide, dapsone, and systemic chemotherapy including melphalan, vincristine, cyclophosphamide, azathioprine, and bortezomib, have all been shown to be effective treatment alternatives. Phototherapy modalities, such as PUVA, narrowband ultraviolet B, and 308-nm excimer lamp, have also shown efficacy in managing cutaneous plasmacytosis.[15,16]

Based on the patient's skin biopsy, deep infiltration of plasma cells was observed. Because UVA has a longer wavelength than NUVB, which can only penetrate the epidermis and superficial dermis, it can penetrate deeper into the dermis and subcutaneous layer, the PUVA was carried out. The mechanisms by which PUVA works include the depletion of langerhans cells, the cross-linking of DNA via psoralen photoadducts, the inhibition of DNA replication, and the immunosuppressive effects on T-lymphocyte function and migration, which lower cytokines and the proliferation and differentiation of plasma cells.[17–19]

In this particular case, the patient underwent a treatment regimen consisting of topical corticosteroids and PUVA therapy, resulting in a gradual reduction in both the size and number of the lesions. Continuous monitoring is being conducted to assess the possibility of systemic involvement. However, further extensive research is required to gain a comprehensive understanding of the underlying mechanisms driving cutaneous plasmacytosis and to establish standardized treatment guidelines for this intriguing dermatological condition.

AUTHOR CONTRIBUTIONS

Thanyathorn Nuchanatanon: Conceptualization; data curation; formal analysis; investigation; methodology; project administration; supervision; writing – review and editing. **Settanan Plangsiri:** Data curation; investigation; writing – original draft. **Teerapong Rattananukrom:** Conceptualization; data curation; formal analysis; investigation; methodology; project administration; supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

There is no conflict of interest to be declared.

DATA AVAILABILITY STATEMENT

All data generated or analyzed during this study are included in this article and its supplementary material files. Further enquiries can be directed to the corresponding author.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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