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Research Article

An Ulcerative Cutaneous Plasmacytosis of the Scalp

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ABSTRACT

Introduction: Cutaneous plasmacytosis (CP) is a rare benign disorder characterized by polyclonal proliferation of plasma cells of unknown etiology, with potential for malignant transformation. Clinical manifestations include reddish-brown macules, papules, plaques, or nodules, and, infrequently, ulcerative lesions resembling cutaneous squamous cell carcinoma (SCC) or similar cutaneous neoplasms. **Case:** Herein we reported a case of ulcerative CP which was initially suspected as SCC in a 77–year–old male. Physical examination revealed an ulcer with well–defined borders, and flat edges, with a granulation tissue base on the scalp which was suggested as SCC. Histopathological features showed infiltration of mature plasma cells in the perivascular area, polyclonal plasma cell population on kappa and lambda chain immunohistochemistry, and hypergammaglobulinemia on protein electrophoresis that supported the diagnosis of CP. **Discussion:** Cutaneous plasmacytosis requires a comprehensive diagnostic evaluation, which typically includes clinical examination, histopathological analysis, immunohistochemistry, ancillary tests such as protein electrophoresis, and negative results from a diagnostic workup for systemic disease. **Conclusion:** The complexity of CP manifestations necessitates a rigorous diagnostic approach, enabling the differentiation of this benign condition from malignancies with similar presentations.
Keywords: Cutaneous plasmacytosis, Hypergammaglobulinemia, Plasma cell.

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INTRODUCTION

Cutaneous plasmacytosis represents an infrequent skin disease characterized by the infiltration of polyclonal plasma cells in the skin and concurrent clonal hypergammaglobulinemia.[1,2] As of 2021, 118 documented cases of CP have been reported globally,[1] with the majority occurring among Asian adult populations, particularly in Japan.[3] Clinical manifestations of CP encompass the appearance of reddish-brown macules, papules, plaques, or nodules distributed across the body and face,[4] occasionally presenting as ulcerative lesions.[4,5] This case report aims to report one rare case of ulcerative CP of the scalp in a 77 year-old-male.

CASE REPORT

A 77-year-old male presented to our Dermatology and Venereology Clinic with a significant complaint of an ulcer on the scalp accompanied by pruritus and pain. The skin lesion first appeared around two years ago as a reddish brown plaque, similar in size to a coin, on the central and posterior vertex. The lesion was prone to bleeding upon scratching, and eventually developed erosion and ulceration. Approximately one year before seeking medical care, the lesion expanded to encompass an area comparable to the size of an adult's palm. The patient currently has no history of systemic disease. On physical examination, the patient exhibited a pain scale of 2/10, normal nutritional status, and displayed alopecia in several areas. Dermatological status revealed an ulcer with well–defined borders, and flat edges, with a granulation tissue base of the scalp (Figure 1). On direct microscopic examination with Gram staining of the ulcers on the scalp, neither Gram-positive nor Gram-negative bacteria were found. On dermoscopy examination found ulcers, blood spots, and polymorphic and irregular blood vessels. Histopathological features showed infiltration of mature plasma cells in the perivascular area (Figure 2), plasma cell population on kappa and lambda chain immunohistochemistry in the ratio of 1:1 indicating polyclonality (Figure 3). Further, protein electrophoresis revealed hypergammaglobulinemia, providing supporting evidence for the diagnosis of CP. The patient was scheduled to undergo wide excision surgery by the Oncology and Plastic Surgery team (Figure 4). Initially, wound management involved conservative measures, including cleansing with 0.9% NaCl solution and applying modern wound dressings with hydrocolloid plaster. Subsequently, topical therapy was implemented employing a combination cream containing silver sulfadiazine and hyaluronic acid, which was continued until the surgical procedure was performed.

DISCUSSION

Cutaneous plasmacytosis is a type of skin disorder that belongs to the cutaneous pseudolymphoma group.[6] It occurs due to the benign proliferation of mature plasma cells and is more prevalent among Asian populations, particularly in Japan, primarily affecting adults.[3] The etiology of CP remains elusive, thought it is believed to arise as a reaction to various stimuli, including trauma, infection, or malignancy.[4,7] A case reported by Lee *et al.*[8] in 1996 even documented a CP case progressing to malignant plasmacytoma on the scalp. The patient in this case report had no history of systemic co-morbidities, malignancy, or infection. However, friction and scratching are suspected triggers for CP in this patient

The classification of plasmacytosis was initially proposed by Watannabe *et al* in 1986, which introduce the term "cutaneous plasmacytosis" to

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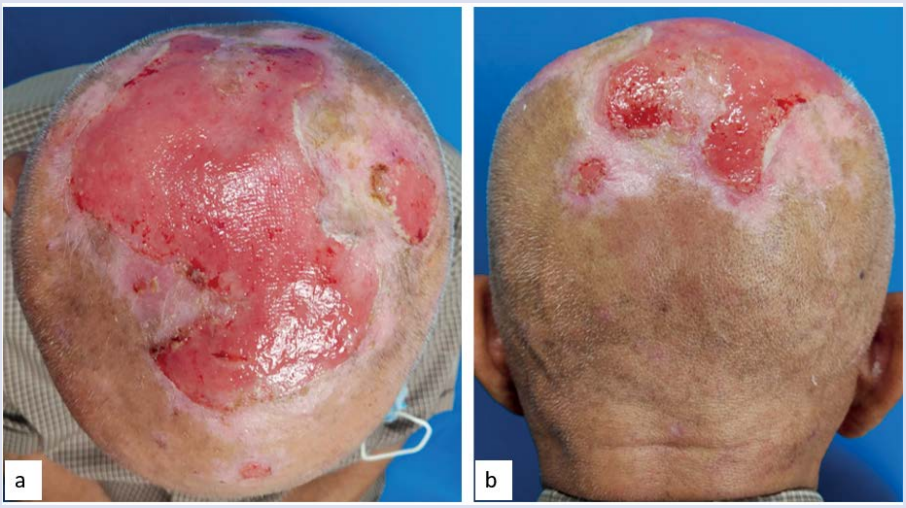

Figure 1: An ulcer with well–defined borders, and flat edges, with a granulation tissue base at (A) central vertex and (B) posterior vertex

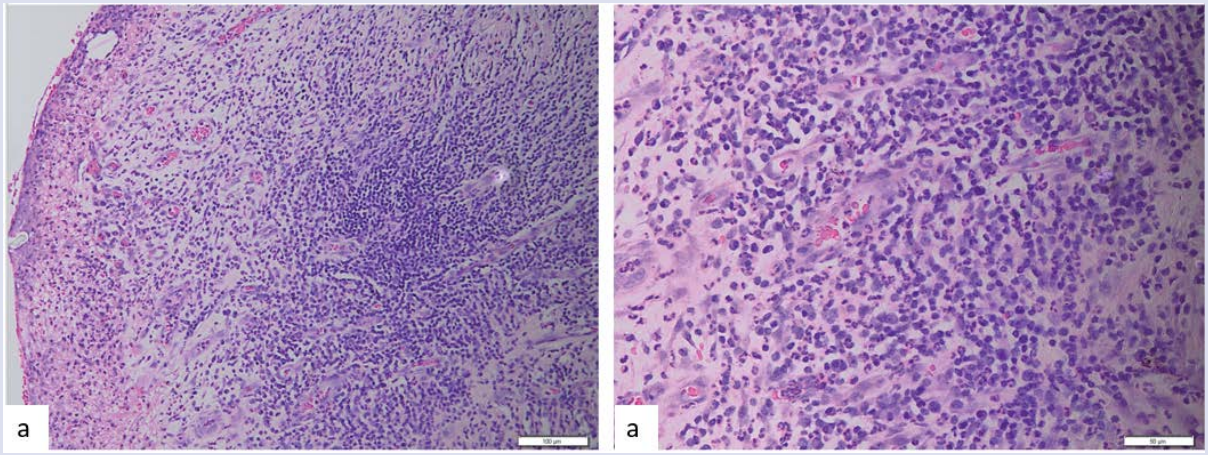

Figure 2: (a) Presence of numerous plasmacytes in the dermis of an ulcer (Hematoxylin & eosin 100X); (b) Perivascular plasma cells infiltrates (Hematoxylin & eosin 200X)

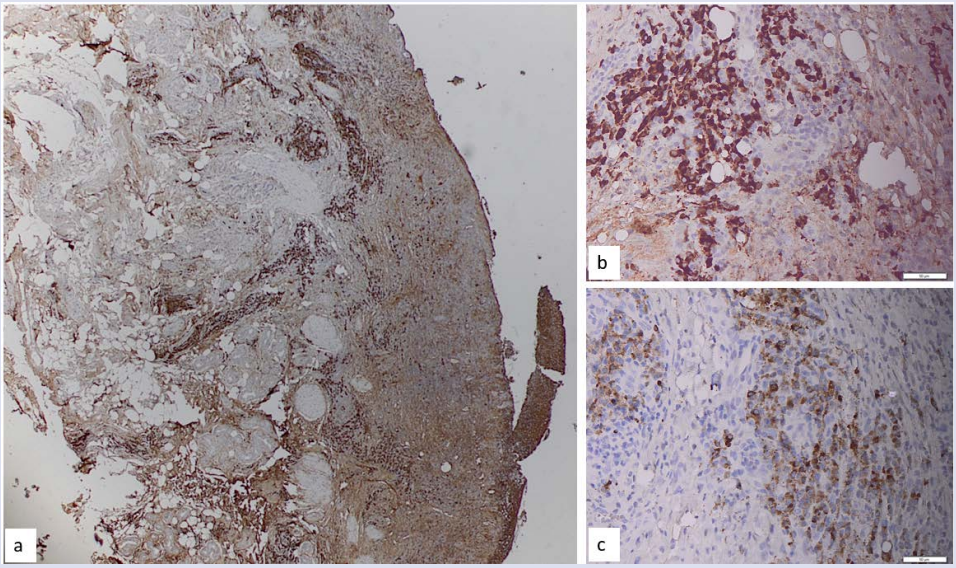

Figure 3: Immunohistochemistry (IHC) shows (a) polyclonal mature plasma cells infiltrate with (b) kappa and (c) lambda light chain (1:1 positivity)

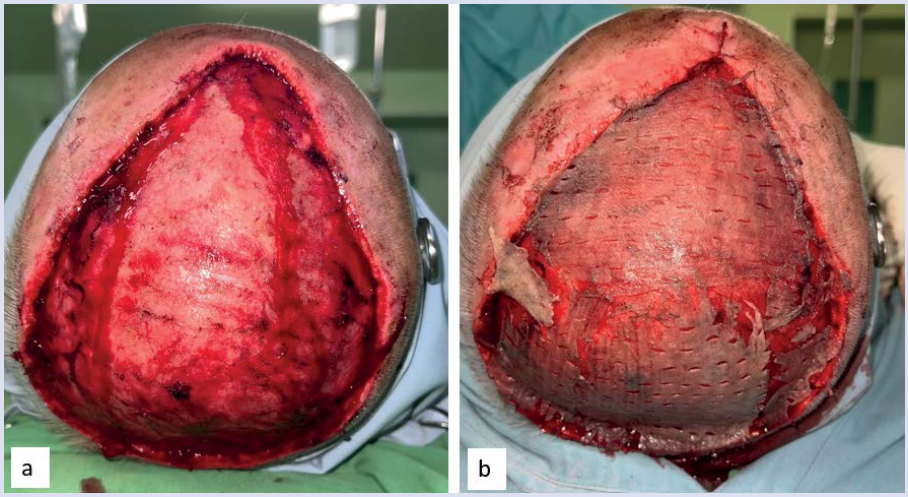

Figure 4: (a) Wide excision of the lesion and (b) closure of the defect

differentiate it from CP with concurent systemic involvement. The presence of systemic involvement in more than two organs is classified as systemic plasmacytosis.[7] Increased plasma cell infiltration in other organs, generally occurring in the bone marrow, lymph nodes, liver, and spleen.[1,5] According to a literature study of plasmacytosis cases, superficial lymphadenopathy (58%) and hepatosplenomegaly (28%) are common indicators of systemic plasmacytosis.[9] However, the patient in this case exhibited neither lymphadenopathy nor hepatosplenomegaly, suggesting the absence of systemic involvement.

Diagnosis of CP relies on a combination of clinical symptoms, histopathological findings, and the exclusion of systemic disease through diagnostic assessments.[5] Clinically, classic CP is typified by the presence of macules, papules, plaques, or reddish-brown nodules, with a predilection for distribution on the body and face.[4,6] In rare instances, these lesions may evolve into ulcers.[4,5] A case reported by Antonio *et al.*[5] found one case of CP with clinical findings of ulcerated plaques on the face. The hallmark histopathological feature of CP is the presence of plasma cell and lymphocyte infiltrates in the perivascular area of the dermis.[5,7] Plasma cells are primarily found in the perivascular dermis, while lymphocytes tend to be concentrated in the middle and lower layers of the dermis.[7] Distinguishing CP from plasmacytoma, a malignant form of plasmacytosis, is imperative.[5] Immunohistochemistry examinations using Kappa and Lambda can aid in this differentiation, revealing the presence of benign plasma cell abnormalities in CP cases, characterized by a polyclonal mature plasma cell infiltrate.[7] Additionally, laboratory findings often reveal hypergammaglobulinemia, observed in 93% of cases.[7,9] In this case, clinical findings manifested as reddish–brown plaques progressing into ulcers, with no history of systemic disease. Physical examination did not reveal any lymphadenopathy or hepatosplenomegaly, confirming the absence of systemic involvement. Histopathological examination revealed mature plasma cell infiltration in the perivascular area, immunohistochemistry demonstrated a kappa and lambda ratio of 1:1, and hypergammaglobulinemia was confirmed by protein electrophoresis, supporting the diagnosis of CP.

At present, there is no standardized treatment regimen for CP, with various therapeutic modalities employed. These include oral, topical, and intralesional corticosteroids, topical tacrolimus, topical psoralen plus ultraviolet A (PUVA), radiotherapy, and photodynamic therapy.[1] In cases of plasmacytosis with large solitary lesions, therapy through radiotherapy or surgery may be considered.[10] The patient in this case had extensive scalp lesions, and the patient's procedure was planned for wide excision surgery by the Oncology and Plastic Surgery team. Initially, conservative wound care was carried out by cleaning the wound with 0.9% NaCl solution and modern wound dressing with hydrocolloid plaster. Later, this therapy was replaced with topical therapy with silver sulfadiazine-hyaluronic acid combination cream until surgery was performed.

CONCLUSION

Cutaneous plasmacytosis is diagnosed through a comprehensive diagnostic evaluation. The occurrence of ulcerative CP, as presented in this case, represents a rare manifestation of this condition. As such, a comprehensive assessment for systemic diseases is imperative before definitively diagnosing CP.

ETHICAL STATEMENT

The publication of images were included in the patient's consent for publication of the case.

CONSENT STATEMENT

The authors certify that they have obtained all appropriate patient consent forms. The patient signed a consent form for the publication of the case details and images.

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CONFLICTS OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

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