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Cutaneous Plasmacytosis: A Rare Encounter

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Abstract **Case Report**

Background: Cutaneous plasmacytosis (CP) is a rare skin condition characterized by the presence of multiple reddish-brown nodules resulting from polyclonal plasma cell proliferation. While it is most commonly reported on the trunk, it can also affect the face and extremities in adults, with a higher prevalence in Asian populations. The underlying cause of CP remains unclear, and there is no established consensus on effective treatment approaches. ***Case report*:** A 58-year-old man with a background of renal failure on hemodialysis seeks advice regarding persistent asymptomatic papules and plaques on the back, neck and trunk which gradually increased over three years. Physical examination revealed multiple discrete infiltrative erythematous to brownish plaques and nodules on the face, neck, trunk and back, with some pustules in certain areas. The skin biopsy showed mature plasmacytic dermal infiltrate requiring immunohistochemical study. The immunohistochemistry showed no light chain restriction, confirming the polyclonal nature of the plasma cells, which supports the diagnosis of plasmacytosis the novelty of the article lies in the first published case of African descent. ***Discussion*:** Cutaneous plasmacytosis is a rare condition of uncertain origin, often indicating a slow-developing systemic disorder. It predominantly affects middle-aged men of Japanese ancestry. The typical manifestations include multiple red-brown papules and plaques with minimal changes to the epidermis. Diagnosis relies on histopathological examination, revealing dense perivascular infiltrates of mature polyclonal plasma cells without atypia. Systemic involvement, termed systemic plasmacytosis, is characterized by cutaneous lesions alongside lymphadenopathy and polyclonal hypergammaglobulinemia. Treatment strategies for cutaneous plasmacytosis remain scarce, lacking a standardized approach.

Keywords: Cutaneous plasmacytosis, rare, African descent.

INTRODUCTION

Cutaneous plasmacytosis is a rare disease of unknown etiology that may represent an indolent systemic process. Most cases have been reported in middle-aged men of Japanese descent [1]. However, there is limited data regarding its occurrence on the African continent. Lesions of cutaneous plasmacytosis typically present as multiple red-brown papules and plaques with little epidermal change. The diagnosis is made by histopathology, which demonstrates dense dermal perivascular infiltrates of mature polyclonal plasma cells without atypia.

REPORT OF A CASE

A 58-year-old male with a background of renal failure presented with the history of persistent asymptomatic papules and plaques on the back, neck and trunk which gradually increased over three years. Physical examination revealed multiple discrete infiltrative erythematous to brownish plaques and nodules on the face, neck, trunk and back, with some pustules in certain areas, The lymph node examination showed no abnormalities, particularly no peripheral lymphadenopathy. Abdominal, cardiovascular, and pulmonary examinations were also normal. The skin biopsy showed mature plasmacytic dermal infiltrate requiring immunohistochemical study. The immunohistochemistry revealed no light chain restriction which goes with the polyclonal nature of the plasma cell infiltrate, the infiltrating plasma cells were positive for CD138, CD4, but negative for CD21, CD117 and CD23.There were no Bence-Jones proteins in the urine. Hemogram, biochemical blood analysis and serum protein electrophoresis were normal, Serological analyses for HIV, syphilis, hepatitis B and C, were all negative. The bone marrow biopsy was not performed following the hematologists' advice.

These clinical and histopathological features, such as the lack of hypergammaglobulinemia, lymphadenopathy, or hepatosplenomegaly, along with the presence of mature polyclonal plasma cell infiltration confined to the skin, are characteristic of isolated benign cutaneous plasmacytosis (PCP).

Therapeutic Management: After confirming the absence of systemic involvement, The patient was prescribed a 2 month regimen of doxycycline, on the follow up the skin lesions are starting to improve.

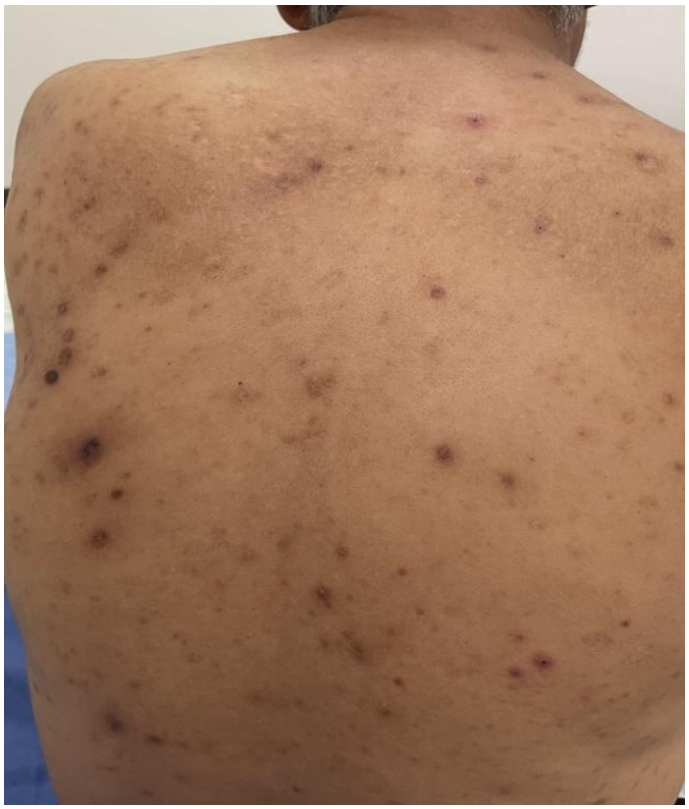

Figure 1: Many brownish, round macules and plaques on the trunk.

DISCUSSION

Cutaneous plasmacytosis (CP) is an uncommon disorder characterized by polyclonal plasmacytosis, primarily affecting the skin. It was initially described by Yashiro in 1976 [2] as "A kind of plasmacytosis" and later termed CP by Kitamura *et al*., [3].

The condition exhibits a higher prevalence in Asia, particularly among the Japanese population. The average age of onset is 37 years, with the typical age range spanning from 20 to 62 years. The presentation typically includes numerous asymptomatic to mildly itchy red-brown papules and plaques with minimal to no epidermal alteration. If systemic involvement occurs, a combination of cutaneous lesions, lymphadenopathy, and polyclonal hypergammaglobulinemia is commonly observed. Apart from lymphadenopathy, other visceral sites of plasmacytic involvement may encompass the bone marrow, lungs, liver, kidneys, or spleen [4].

Diagnosing cutaneous plasmacytosis usually involves histopathological examination, often supported by immunohistochemical staining of a biopsy sample. Histopathological analysis typically reveals dense perivascular infiltration of plasma cells in the dermis without notable atypia. In cutaneous plasmacytosis, the plasma cell infiltrates are polyclonal, whereas a monoclonal infiltration may indicate marginal-zone B-cell lymphoma. Additional histopathological findings may include elevated levels of mast cells along with varying numbers of lymphocytes and histiocytes [5].

Laboratory findings linked with systemic plasmacytosis comprise polyclonal hypergammaglobulinemia, with a kappa: lambda ratio falling within the normal range.

Despite being recognized for several decades; the precise etiology and pathogenesis of CP remain elusive. However, it is suggested that dysregulated production of IL-6 may play a pivotal role in its pathogenesis. Some researchers speculate that CP could arise from mutations in genes encoding signaling molecules crucial in plasma cell regulation. Others suggest a potential connection with elevated levels of IL-6, which is known for its role in driving the terminal differentiation of B cells into plasma cells [6].

Managing cutaneous plasmacytosis poses challenges due to its unpredictable response to treatment. In addressing cutaneous lesions, intralesional corticosteroids have demonstrated moderate effectiveness. Other treatments investigated for cutaneous lesions encompass topical calcineurin inhibitors, phototherapy methods such as psoralen with ultraviolet A light, radiotherapy, and intralesional interferon-gamma [7-9]. Doxycycline has been proposed in some cases due to its anti-inflammatory and immunomodulatory properties, which may help reduce the skin lesions associated with CP [10].

CONCLUSION

In conclusion, the description of the first documented case to our knowledge of cutaneous plasmacytosis in Africa sheds light on the importance of recognizing and characterizing this rare entity in diverse geographical settings. This case underscores the diagnostic and therapeutic challenges associated with this condition, as well as the need for further research to better understand its pathogenesis and optimal management. By expanding the knowledge base on cutaneous plasmacytosis, this case report contributes to improving the care of patients with this rare disease, particularly in regions where its prevalence may be under-recognized.

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