

# Cutaneous plasmacytosis: A rare entity with unique presentation

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## Abstract

Primary cutaneous plasmacytosis is a rare cutaneous disorder with extensive cutaneous plaques/papules mainly on the trunk and face. Cases have mostly been documented from Japan. We present here a rare case of cutaneous plasmacytosis from India of Mongolian descent. This 50-year-old female from Mizoram had extensive maculo-papular violaceous plaques distributed on the face, axillae, trunk and lower extremities. Initial and repeat skin biopsy revealed dense perivascular and periadnexal mature plasma cells. She also had lymphadenopathy. Serum protein electrophoresis did not reveal any M band and the Bence Jones protein was negative in urine. The patient had multiple superficial lymph nodes and a biopsy from the cervical lymph node showed effacement of normal nodal architecture by sheets of plasma cells. Immuno histochemistry was done from both skin and lymph node biopsies. The kappa and lambda tight chains were not restricted; there by proving the polyclonal nature of the plasma cells. The novelty of the case lies in its classical clinical presentation with histopathological documentation.

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## Introduction

Primary cutaneous plasmacytosis is a rare cutaneous disorder with extensive cutaneous plaques/papules mainly on the trunk and face. This entity is thought to be a reactive process of unknown etiology.<sup>1</sup> The skin lesions may be associated with variable extracutaneous involvement.<sup>2</sup> Cases have mostly been documented from Japan.<sup>3</sup>

We present here a very rare case of cutaneous plasmacytosis from India in a 53-year-old female of Mongolian descent.

## Case Report

A 50-year-old female from Mizoram (north-eastern part of India) and of Mongolian ethnicity presented with multiple violaceous plaque lesions of variable size and shapes all over the body, predominantly over trunk, face and neck. Examination revealed reddish brown to purple colored plaques of sizes varying from 3-6 cm in diameter. The shapes were varied, viz., circular, elongated, annular, aciform coalescing at places forming reticulated patches [Figure 1]. Lesions were found predominantly over the trunk, face and neck [Figure 2], axillae

and back. No hepato splenomegaly was found. Patient was found to be anaemic. A skin biopsy was performed and initial blood tests were ordered. Her Hemoglobin was found to be 7.3 gm/dl and the ESR was 150/hr. Blood for HIV antibodies (1 and 2) were negative and VDRL was non reactive. The histopathology examination of the skin biopsy revealed the following: The epidermis was unremarkable. The dermis shows moderately dense aggregates of plasma cells in the superficial and deep perivascular areas [Figure 3]. These were admixed with variable number of lymphocytes and histiocytes [Figure 4].

A peripheral smear examined revealed extensive rouleaux formation. Other biochemical parameters were as follows - BUN: 26.8 mg/dl, Creatinine 3.4 mg/dl, total protein was 12.6 g/dl, albumin was 2.6 gm/dl, globulin was 10g/dl, total urinary protein was 15931 mg/dl, LDH was 361 U/L. Serum total IgG was 41 g/dl.

Serum protein electrophoresis did not reveal any M band and the Bence Jones protein was negative in urine.

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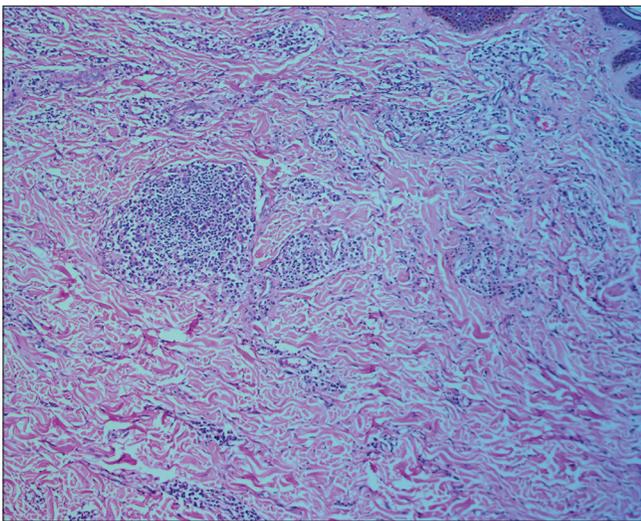
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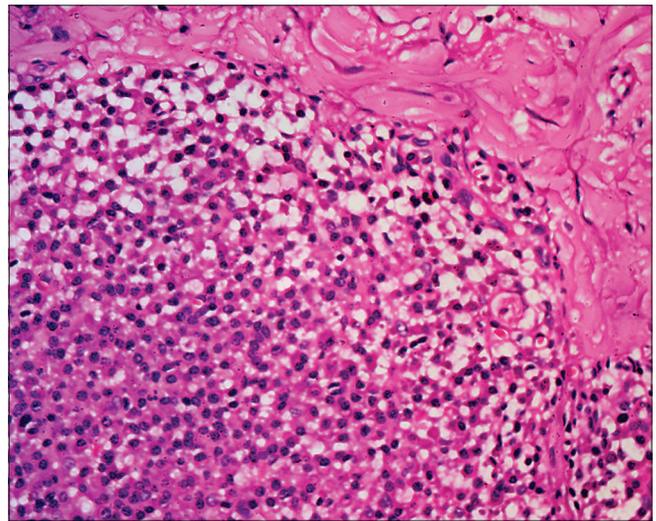
**Figure 1:** Multiple violaceous coloured plaques of variable sizes and shapes over back



**Figure 2:** Violaceous plaques over face and neck in the same lady



**Figure 3:** Lymphoplasmacytic infiltrate in the dermis (H and E, ×100)



**Figure 4:** Abundant plasma cells and lymphocytes in dermal infiltrate (H and E, ×400)

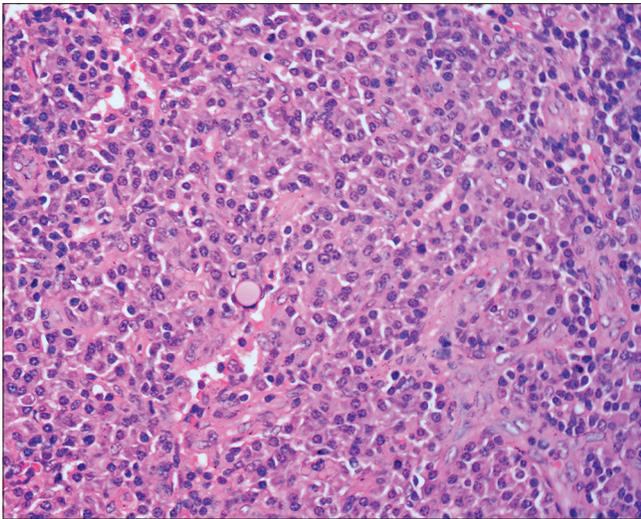
The patient also had multiple superficial lymph nodes in the cervical, axillary and inguinal region. Biopsy from the cervical lymph node showed effacement of normal nodal architecture by sheets of plasma cells [Figure 5].

Immunohistochemistry was done for kappa and lambda light chains to rule out monoclonality of plasma cells from the lymph node biopsy. The kappa and lambda tight chains were not

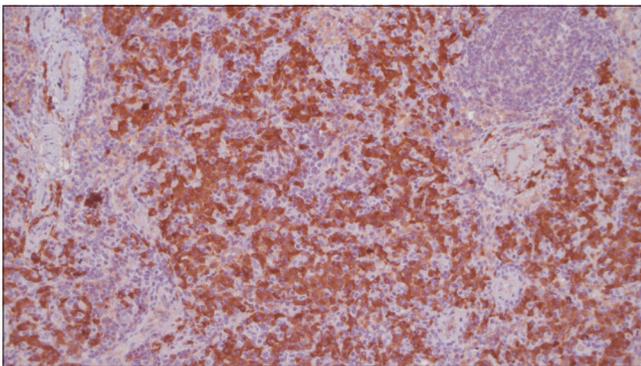
restricted, thereby proving the polyclonal nature of the plasma cells [Figure 6].

### Discussion

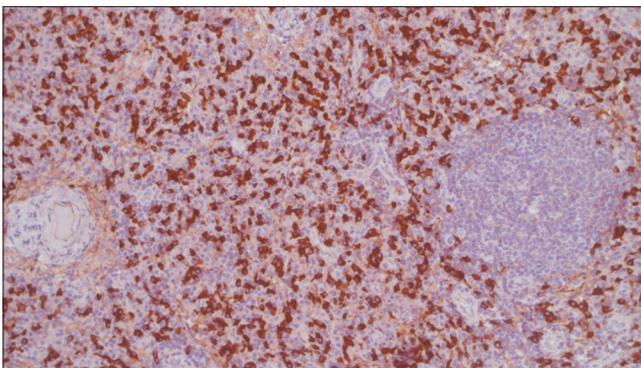
Cutaneous and systemic plasmacytosis is a rare reactive lympho-plasmacytic disorder. It was first described in Asia by Yashiro in 1976 who described it as a type of plasmacytosis. Kitamura



**Figure 5:** Sheets of plasma cells in lymph node (H and E, ×400)



**Figure 6a:** Positive IHC for kappa light chain



**Figure 6b:** Positive IHC for lambda light chain

further characterised it in 1980. Since then it has been described in Asian patients, specially amongst the Japanese.<sup>4</sup> As most patients of cutaneous plasmacytosis are found to have some form of systemic symptoms,<sup>5</sup> some authors prefer the term “cutaneous and systemic plasmacytosis” to account for the extracutaneous involvement.<sup>6</sup>

The cutaneous plasma cell infiltrate are polyclonal and the most common extracutaneous involvement is lymphadenopathy.

Clinical manifestations include fever, anaemia and hypergammaglobulinaemia. The cutaneous presentation is very

characteristic comprising of multiple reddish-brown infiltrated macules, plaques and nodules, chiefly on the trunk.<sup>2</sup>

The etiopathogenesis is poorly understood. Dysregulated production of IL-6 which is a cytokine that induces B - cell proliferation and differentiation to plasma cells which in turn causes immunoglobulin secretion and angiogenesis is thought to play a role. Genetic polymorphism in the IL-6 gene may possibly explain the propensity of the disease to manifest in particular geographic regions.<sup>7,8</sup>

Recently a similar case of primary cutaneous plasmacytosis has been reported from India.<sup>9</sup> We are not aware of any such case where Hidradenitis Suppurativa has been a manifestation of primary cutaneous plasmacytosis as reported in the case. This is to our mind, certainly an unusual manifestation.

Histologic features include a moderately dense infiltrate of mature plasma cells in perivascular areas of both superficial and deep dermis. Admixture of lymphocytes may be present and lymphoid follicles with reactive germinal centres can also be seen.

Light chain restriction is absent which goes with the polyclonal nature of the plasma cell infiltrate.<sup>10</sup>

Approximately 58% of cases of cutaneous plasmacytosis have associated lymphadenopathy which forms the commonest extracutaneous manifestation. Liver, spleen, lungs and kidney are less frequently affected organs.<sup>1,11</sup> Our patient had no history of recurrent infections, chronic inflammatory disease or neoplasm and tested negative for autoimmune markers. Although creatinine was raised in our patient, the kidneys could not be assessed as the patient refused kidney biopsy.

Most cases of plasmacytosis run a benign indolent course without spontaneous remissions. There are reports of plasmacytosis developing malignant lymphoma. However the rate of transformation to malignant neoplasm is quite low in Asian literature.<sup>2</sup>

Cases are recalcitrant to treatment. Treatment modalities like topical and systemic steroid, antibiotics and systemic chemotherapy have shown to have some beneficial effects. Topical 0.1% tacrolimus, thalidomide, photodynamic therapy, combining corticosteoid with cyclophosphamide, topical PUVA, radiotherapy have been tried with variable outcome.<sup>12</sup>

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

#### References

1. Chantachaeng W, Chularojanamontri L. Cutaneous plasmacytosis: A case report and review of pulmonary findings. *Dermatol Reports* 2011;3:e39.

2. Lee TG, Jeong WS, Moon SH, Hwangbo H, Lee SK, Lee DR, *et al.* Cutaneous and systemic plasmacytosis associated with renal amyloidosis. *Ann Dermatol* 2015;27:759-62.
3. Uhara H, Saida T, Ikegawa S, Yamazaki Y, Mikoshiba H, Nijoh S, *et al.* Primary cutaneous plasmacytosis: Report of three cases and review of the literature. *Dermatology* 1994;189:251-5.
4. Hjjira N, Baba N, Boudhas A, Al Bouzidi A, Boui M. Cutaneous plasmacytosis: Report of a Moroccan case. *J Dermatol Dermatolog Surg* 2014;18:27-30.
5. Tada Y, Komine M, Suzuki S, Kikuchi K, Sasaki M, Kaneko N, *et al.* Plasmacytosis: Systemic or cutaneous, are they distinct? *Acta Derm Venereol* 2000;80:233-5.
6. Honda R, Cerroni L, Tanikawa A, Ebihara T, Amagai M, Ishiko A. Cutaneous plasmacytosis: Report of 6 cases with or without systemic involvement. *J Am Acad Dermatol* 2013;68:978-85.
7. Kodama A, Tani M, Hori K, Tozuka T, Matsui T, Ito M, *et al.* Systemic and cutaneous plasmacytosis with multiple skin lesions and polyclonal hypergammaglobulinaemia: Significant serum interleukin-6 levels. *Br J Dermatol* 1992;127:49-53.
8. Fang S, Shan K, Chen AJ. Cutaneous and systemic plasmacytosis on the face: Effective treatment of a case using thalidomide. *Oncol Lett* 2016;11:1923-5.
9. Goyal T, Varshney A, Zavar V, Sharma V. Primary cutaneous plasmacytosis: Masquerading as hidradenitis suppurativa. *Indian J Dermatol* 2016;61:213-5.
10. Amin HM, McLaughlin P, Rutherford CJ, Abruzzo LV, Jones D. Cutaneous and systemic plasmacytosis in a patient of Asian descent living in the United States. *Am J Dermatopathol* 2002;24:241-5.
11. Yang YW, Hu S, Kuo TT, Hong HS. Cutaneous plasmacytosis-Report of a case. *Dermatol Sin* 2003;21:97-101.
12. Wong KF, Chan JK, Li LP, Yau TK, Lee AW. Primary cutaneous plasmacytoma – Report of two cases and review of the literature. *Am J Dermatopathol* 1994;16:392-7.