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Case Report

Dermatology Section

Primary Cutaneous Plasmacytosis with Calcinosis: A Rare Case Report

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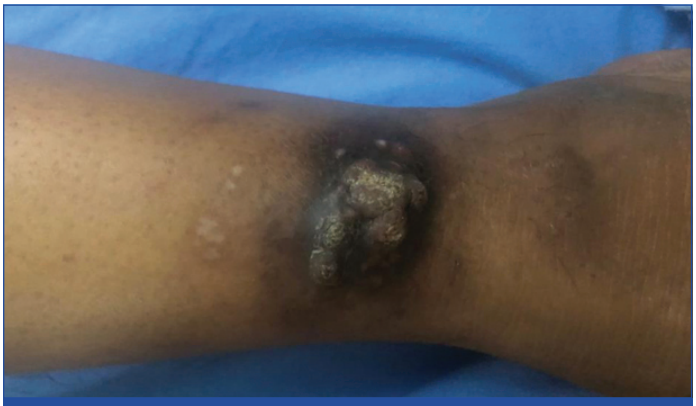
ABSTRACT

Primary cutaneous plasmacytosis is a rare disorder. Cases have been mostly reported from Japan. Here, a rare case of cutaneous plasmacytosis with calcium deposits was reported from southern part of India. A 47-year-old male patient presented with solitary asymptomatic nodule on the left leg present for two decades. Skin biopsy showed sheets of plasma cells in mid-dermis with bony trabeculae extending upto subcutis which was unremarkable. Serum electrophoresis did not reveal any M band and Bence Jones protein was absent in urine. Radiograph of the left leg showed focal lesion with calcific areas in soft tissue shadow with probability of calcification. Novelty of the case lies in its rare clinical presentation in this population with histopathological documentation.

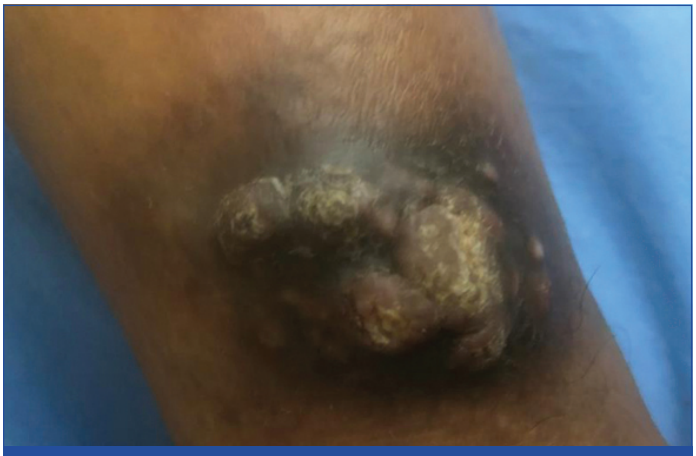
Keywords: Plasma cells, Skin biopsy, Skin nodule, Subcutaneous calcification

CASE REPORT

A 47-year-old male patient presented to Dermatology Outpatient Department with hyperpigmented plaque and nodule measuring 6×7 cm present over lateral aspect of left leg 6 cm above the lateral malleoli [Table/Fig-1,2]. The lesion was asymptomatic and present since two decades with no history of significant variation in the size. There was no evidence of other lesions anywhere else in the body. It was not associated with any systemic complaints. There was no history of similar complaints from the parents and siblings. The index patient is married with two children. On general physical examination, the patient was of a morderate built and nourishment, no pallor, no cyanosis, no clubbing and no pedal oedema. There was no evidence of lymphadenopathy or hepatosplenomegaly.



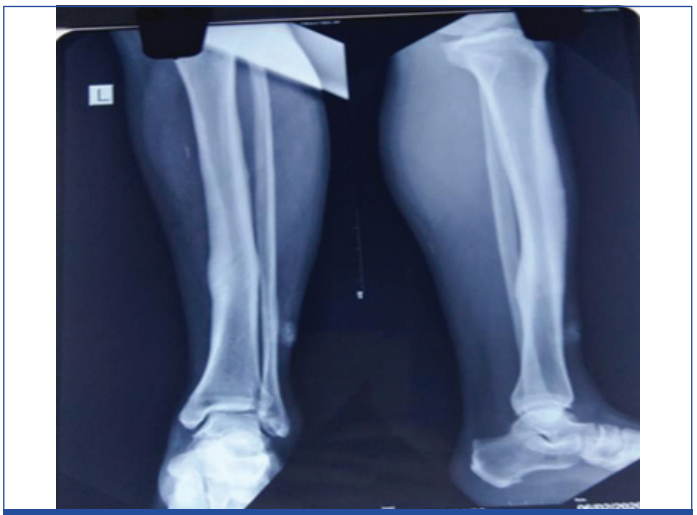
[Table/Fig-1]: Hyperpigmented plaque (6×7 cm) seen on the lower third of leg, lateral aspect.



[Table/Fig-2]: Hyperkeratotic plaque with scales.

Differential diagnoses of calcinosis cutis, sarcoidosis, secondary deposits from unknown primary tumor was thought of and the patient was subjected to routine blood investigations, skin biopsy and radiography.

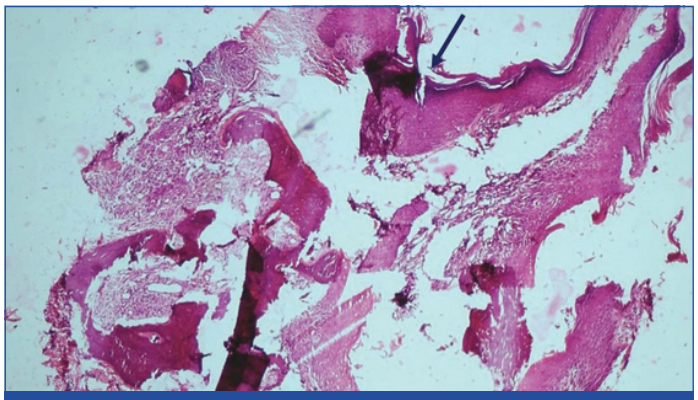
Routine blood investigations revealed haemogloblin (Hb) -16.6 g/dL Erythrocyte Sedimentation Rate (ESR) 440 mm/hr. Peripheral smear revealed normocytic normochromic picture. Renal function test and liver function tests were within normal limits. Urine routine and microscopic examination was normal. Human immunodeficiency virus, hepatitis B, rapid plasma reagin test were non reactive. Radiograph of left leg Anteroposterior (AP) and lateral view showed focal lesion with calcific areas in the soft tissue shadow of the leg with probability of calcification or hetrotrophic ossification [Table/Fig-3]. Patient was subjected to chest radiograph radiograph of skull and lumbar spine, both AP and lateral view, which were normal.



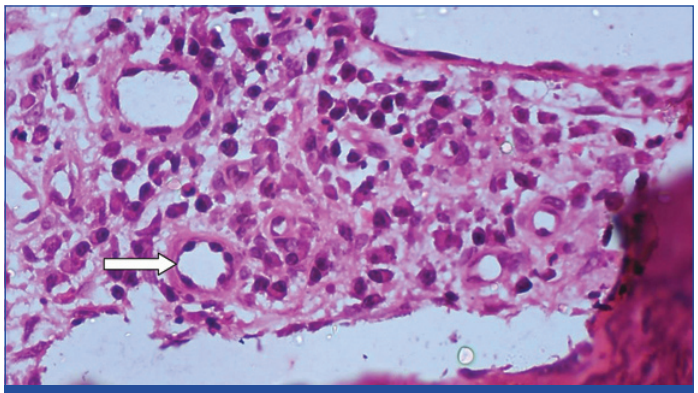
[Table/Fig-3]: X-ray left foot and leg showing focal lesion with calcific areas in soft tissue shadow of leg. (lower one third), AP and Lateral view.

A skin biopsy of 3 mm thickness taken from the plaque showed irregular acanthosis in the epidermis. Dermis showed trabecular bone tissue with localised aggregates of plasma cells in sheets and clusters which were oval with eccentrically placed nucleus and cartwheel shaped chromatin. Bony trabeculae is seen up to subcutaneous tissue and appears unremarkable [Table/Fig-4,5].

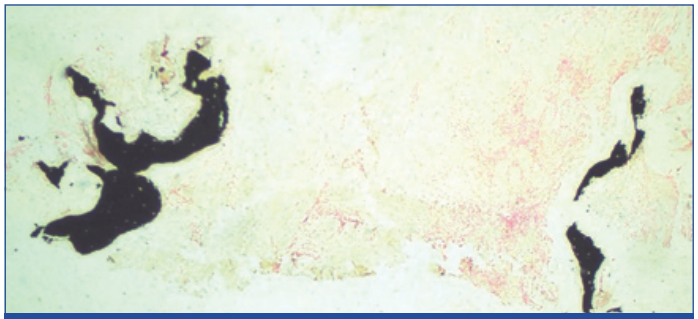
The histopathological study with Von Kossa staining highlighted the calcium deposits and bony trabeculae [Table/Fig-6-8].



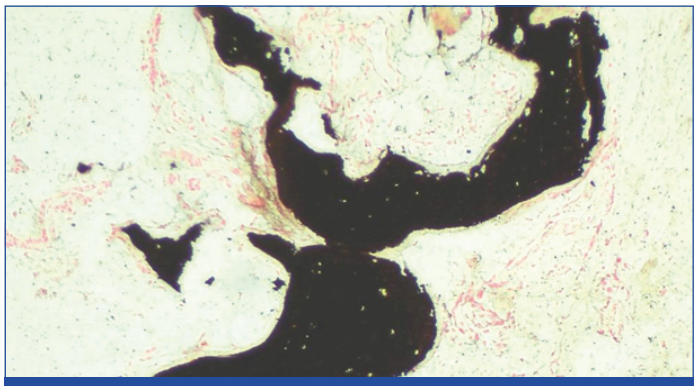
[Table/Fig-4]: Epidermis with irregular acanthosis (arrow). Dermis shows trabecular bone with aggregates of plasma cells (Magnification- 100x) (H&E).



[Table/Fig-5]: Abundant plasma cells with eccentrically placed nucleus (arrow), cartwheel chromatin and dense eosinophilic cytoplasm (H&E) (Magnification- 400x).

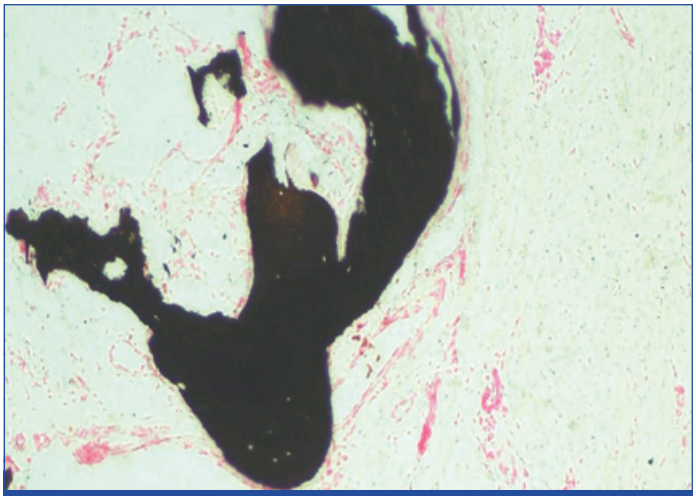


[Table/Fig-6]: Von kossa stain highlighting bony trabaculae extending upto subcutis. (Magnification- 10x)



[Table/Fig-7]: Von kossa stain showing bone. (calcium deposists highlighted) (Magnification-40x).

Patient was advised to apply topical halobetasol 0.05% cream in the morning and fusidic acid 2% cream at night for a period of two weeks. Oral antibiotics containing amoxicillin 500 mg and clavulinic acid 125 mg was prescribed for a week twice daily. At two weeks follow-up the lesion showed partial resolution in size and thickness. Further follow-up after one year done through a telephonic conversation showed stabilisation of the lesion.



[Table/Fig-8]: Calcium deposits enlightened with von kossa; extending upto subcutis (Magnification- 40x).

DISCUSSION

Primary cutaneous plasmacytosis is a rare cutaneous disorder presenting with multiple papules and plaques distributed over trunk and face. The skin lesions may be associated with variable extracutaneous involvement [1]. It is usually diagnosed based on clinical and histopathological correlation. It was first described in Asia by Yashiro in 1976. This condition was described specifically amongst Japanese population [2]. The cutaneous plasma cell infiltrate is polyclonal and the most common extracutaneous involvement is lymphadenopathy.

The cutaneous presentation of plasmacytosis is characteristically multiple reddish-brown infiltrated macules, plaques and nodules, present in mainly on the trunk [2]. However, the lesion in this patient was localised to a single plaque on the left leg without extracutaneous involvement. This is to our knowledge a unique presentation of primary cutaneous plasmacytosis in a patient of south Indian ethnicity.

The aetiopathogenesis is not clearly established. 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. Unfortunately, the IL-6 level estimation was not done in present patient. Genetic polymorphism in IL-6 gene may possibly explain the propensity of the disease to manifest in particular geographic regions [3,4].

Recently, a case of primary cutaneous plasmacytosis was reported from our subcontinent with mast cell infiltration [5]. This case is unique with focal bony infiltration along with primary cutaneous plasmacytosis without lymphadenopathy and systemic symptoms.

Histopathologically, cutaneous plasmacytosis is marked by dense superficial and deep perivascular and periappendageal infiltrate composed mainly of plasma cells along with few lymphocytes and histiocytes. Plasma cells are mature without any evidence of atypia. Other less commonly described features include perineural distribution of plasma cells and lymphoid follicles with reactive germinal centers [6].

The most common extra cutaneous involvement is lymphadenopathy which accounts to approximately 58%. The other less frequently affected organs are liver, kidney, spleen and lungs [7,8]. Present patient had lesions only on left leg which was completely asymptomatic and present since two decades. Liver function test and renal function test were within normal limits. Chest X-ray anterior posterior view was unremarkable.

Most cases of plasmacytosis run a benign indolent course without spontaneous remissions. There is a report of plasmacytosis developing into malignant lymphoma [9]. However, the rate of transformation to malignant neoplasm is quite low in Asian literature [2].

Cutaneous plasmacytosis has a chronic benign course with an overall favourable prognosis. There is no standard effective treatment for this condition, and different treatment options have been tried with limited success [10]. Treatment modalities like topical and systemic steroid, antibiotics and systemic chemotherapy have shown to have some beneficial effects. The patient in present study received topical halobetasol and fusidic acid creams, along with oral amoxclav and had partial response to treatment. Topical 0.1% tacrolimus, thalidomide, photodynamic therapy, combining corticosteroid with cyclophosphamide, topical PUVA and radiotherapy have been tried with variable outcomes [11].

CONCLUSION(S)

Primary cutaneous plasmacytosis is a rare disorder. However it should be considered among the differential diagnoses for long standing nodular and plaque lesions. An incisional biopsy should be done routinely in all patients having long standing lesions.

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