Cutaneous plasmacytosis: a case report and review of pulmonary findings

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Abstract

Primary cutaneous plasmacytosis is an uncommon cutaneous disorder with multiple cutaneous nodules and plaques mainly on face and trunk. This entity is thought to be a reactive process with unknown etiology. Pulmonary involvement could be found as a part of systemic plasmacytosis; however, cutaneous plasmacytosis was also reported with other pulmonary disorders. This report presents the case of cutaneous plasmacytosis and the review of pulmonary findings reported in plasmacytosis.

Introduction

The primary cutaneous plasma cell disorders can range from malignant to benign plasma cell neoplasms. The malignant conditions are neoplastic diseases having monoclonal proliferations, rapid progression and fatal outcome while the benign plasma cell disorders usually show polyclonality, chronicity and benign process, including plasmacytosis. Some authors stated that cutaneous and systemic plasmacytosis should be the same entity because systemic involvement has not been found in the former due to a lack of sufficient investigations. In this report, the authors present a case of cutaneous plasmacytosis, which is a rare reactive skin condition and also a review pulmonary findings in this disease.

Case Report

A 41-year-old male presented with the history of persistent asymptomatic papules and plaques on face, neck and trunk which gradually increased over two years (Figure 1). Physical examination revealed multiple discrete infiltrative erythematous to brownish plaques and nodules on face, neck, trunk and back. The skin biopsy showed superficial and deep perivascular and periadnexal infiltrations with mature plasma cells, admixed with some lymphocytes and histiocytes with no atypicality (Figure 2). The immunohistochemistry revealed no light chain restriction.

Basic chemical laboratory testing was within normal limits. Serology for anti-human immunodeficiency virus, anti-nuclear antibody and syphilis were negative. Urine Bence Jones protein, β2-microglobulin, serum free light chain analysis and serum protein electrophoresis were all within normal limits. Bone marrow biopsy resulted in moderately hypercellularity with normal maturation. However, a speculated lung nodule with cavity 1.7 cm in diameter was seen at right upper lobe from chest X-ray. Three consecutive sputum examinations were negative for acid fast bacilli. With previous reports of pulmonary involvement in plasmacytosis, computer tomography (CT) scan of the chest was done and revealed a group of cavitory pulmonary nodules with nearby tree-in-bud pattern, suggestive of tuberculosis rather than malignancy. Finally, the diagnosis of pulmonary tuberculosis was confirmed by bronchoalveolar fluid culture and lung biopsy.

After successful course of 6-month anti-tuberculosis regimen, there was a resolution of pulmonary lesions but skin lesions still progressed gradually. Intravenous steroid injection and tacrolimus ointment application were commenced but did not show promising results. Then, the patient decided to have a scheduled follow-up visits.

Primary cutaneous plasmacytosis or cutaneous plasmacytosis was thought to be a reactive process with unknown etiology and characterized by disseminated, asymptomatic reddish-brown macules, plaques and nodules mainly on upper trunk and face. Classification for cutaneous plasmacytosis was coined by Watanabe et al. in 1986.

Cutaneous plasmacytosis without pulmonary involvement by definition, had been reported with some abnormal pulmonary findings such as lung carcinoma and lung nodules. A primary plasmacytosis affecting more than two organs was classified as systemic plasmacytosis. The common organ involvement were superficial lymphadenopathy (38%) and hepatosplenomegaly (28%), respectively. Pulmonary findings of systemic and cutaneous plasmacytosis were demonstrated as Table 1. As far as the author’s review, there were two cases which had pulmonary granuloma and plasmacytosis (case No. 4, 5).

Uhara et al. reviewed that the most characteristic histopathologic change of plasmacytosis was perivascular and periadnexal patchy infiltration of matured typical plasma cells with some lymphocytes and histiocytes. Immunohistochemistry was found to be polyclonal IgG-positive predominated. Other important differential diagnosis includes syphilis, Lyme disease, systemic lupus erythematosus and plasmacytoma.

The extensive investigations of our patient showed no distinctive causes of plasma cell proliferation. Firstly, the authors believed that cutaneous plasmacytosis might be a reactive process of pulmonary tuberculosis. However, the skin lesions did not resolve after successful course of anti-tuberculosis drugs. As a result, pulmonary tuberculosis, which occurs com-

[Figure 1. Multiple discrete infiltrative erythematous to brownish papules and plaques on chest wall and trunk.]

[Figure 2. Perivascular and periadnexal infiltrations predominantly with mature plasma cells, admixed with lymphocytes and histiocytes. No atypical plasma cells were observed.]
Table 1. Clinical features of systemic and cutaneous plasmacytosis patients with abnormal pulmonary findings.

| Patient | Age / Sex | Location of lesions | Symptoms | Radiographic and pathologic findings of lung | Diagnosis | Treatments and outcome |
|---------|-----------|---------------------|----------|---------------------------------------------|-----------|------------------------|
| 1.      | 49/F      | Korean              | Fatigue, shortness of breath and dry cough | CT: innumerable tiny perivascular nodules Biopsy: mature plasma cells infiltrations | Systemic plasmacytosis (cutaneous, lymphadenopathy and lungs) | 1. CHOP regimen: partial response 2. Anti CD-20: no response 3. Prednisolone (1mg/kg/D): reduction in pulmonary symptoms and lymphadenopathy but no improvement of cutaneous lesions |
| 2.      | 62/F      | Japanese            | Dyspnea on exertion, wheezing and cough | CXR: reticulonodular both lower lungs Biopsy: follicle formations with infiltration of plasma cells and lymphocytes, alveolar wall fibrosis found | Systemic plasmacytosis (cutaneous, lymphadenopathy and lungs) | Died due to respiratory failure despite prednisolone therapy and various kinds of treatments |
| 3.      | 54/M      | Japanese            | Fatigue | CT: ground-glass attenuation with reticular infiltration in middle and lower lobes Biopsy: thickening of alveolar septum with marked infiltration of lymphoplasmacytic cells | Systemic plasmacytosis (cutaneous, lymphadenopathy and lung) | Prednisolone (15mg/D): partial response |
| 4.      | 67/M      | Chinese             | Asymptomatic, history of treated pulmonary tuberculosis | CT: a foci of ground-glass attenuation with calcified granuloma, multiple axillary and intrathoracic lymphadenopathy Biopsy: not done | Systemic plasmacytosis (cutaneous, lymphadenopathy and lung) | N/A |
| 5.      | 35/F      | Philippino          | Fatigue | CT: 2 small pulmonary nodules Biopsy: not done (thought to represent granuloma) | Cutaneous plasmacytosis with pulmonary nodules | 3 years follow-up without any treatment: stable and asymptomatic |
| 6.      | 55/M      | Caucasian           | Fever, malaise and significant weight loss. History of anal chronic ulcer | CT: pulmonary mass of left lingular lobe with multiple lymph nodes Biopsy: squamous cell carcinoma | Cutaneous plasmacytosis with lung and anal carcinomas | Died from respiratory and circulatory failure |
| 7.      | 35/M      | Thai                | Asymptomatic | CT: cavitary pulmonary nodules and centriflobular nodules at RUL Biopsy: necrotizing granulomatous inflammation | Cutaneous plasmacytosis with pulmonary tuberculosis | Resolved pulmonary lesions after anti-tuberculosis therapy but persistent cutaneous lesions despite of intralesional steroid and topical tacrolimus |

CT: computer tomography; CXR, chest X rays; CHOP, cyclophosphamide, doxorubicin, vincristine, prednisolone.

Conclusions

In summary, the authors describe the rare case of cutaneous plasmacytosis which is diagnosed at the same time of pulmonary tuberculosis. Data from more patients are required in order to conclude that plasmacytosis is a reactive process or co-incidence of tuberculosis. However, lung is another organ that should be taken into consideration for sufficient investigation of plasmacytosis.

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Case Report

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