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Case Report

Cutaneous and Systemic Plasmacytosis in a Case of Rib Pain and Anemia

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Keywords

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Abstract

A 67-year-old woman was admitted to our department presenting with rash, rib pain and anemia. The patient had had a rash for 20 years, and was previously diagnosed with multiple myeloma at her local hospital. The patient doubted both the diagnosis of multiple myeloma and the effectiveness of the treatment. We discovered that this patient had in fact been misdiagnosed for multiple myeloma, and then diagnosed her as having cutaneous and systemic plasmacytosis (CSP) by skin biopsies.

INTRODUCTION

Cutaneous and systemic plasmacytosis (CSP) is a rare condition characterized by a clinical of cutaneous lesions and polyclonal hypergammaglobulinemia. The condition is described as primarily affecting middle-aged Asian patients [1]. The etiology of CSP is unknown,but current evidence suggests that deregulated production of interleukin(IL)-6 plays an important role in its pathogenesis [2,3]. The differenial diagnosis of CSP includes the multicentric plasma cell variant of Castleman disease(MPCD) [4] and idiopathic plasmacytic lymphadenopathy(IPI). CSP defies diagnosis with biopsy. Skin biopsy findings characteristically show a dermal infiltrate composed of mature plasma cells without atypia and variable numbers of admixed lymphocytes and histiocytes. The patient had been misdiagnosed as having multiple myeloma in a local hospital.

CASE REPORT

A 67-year-old woman was admitted to our department because of rash, rib pain, and anemia. The patient had had a rash for 20 years, and it had initially presented on her neck and then spread to her trunk and legs (Figures 1). The whole-body rash consisted of scattered brown and dark red spots of various sizes that did not fade with pressure. The rash caused no discomfort. The patient was diagnosed as having plasma cell granuloma by skin biopsy and was treated with prednisone at 40 mg/day for one month, which reduced her rash color, but the extent of the rash increased. Medications were then discontinued and the patient was put on observation until 2006. After experiencing rib pain for a year, the patient admitted herself to her local hospital in 2006. Her complete blood cell count showed anemia, reduced blood albumin and notably elevated globulin levels. Serum protein electrophoresis revealed markedly elevated gamma globulin levels. X-ray showed bone destruction, and ECT showed a concentration of dots on the ninth rib of her right side, radioactive distribution enhancements to the left ankle, and distributed radioactivity in the rest of her bones. Bone biopsy showed an elevated level of diffuse plasma cells. Multiple myeloma (MM) was diagnosed, and the patient was given 6 cycles of thalidomide and dexamethasone. The bone pain showed no significant improvement, and the rash spread.

The patient doubted her initial diagnosis of multiple myeloma and the effectiveness of the treatment, so she came to our hospital for a second opinion in 2007. On examination, the patient had an anemic appearance, and the painless rash consisted of multiple 0.5- to 2.0-cm-diameter brownish-red markings that were disseminated symmetrically on the neck, chest, back, abdomen, and legs (Figures 1). Bilateral anterior cervical, axillary and inguinal reach lymph nodes were enlarged. The patient did not have any bone tenderness.

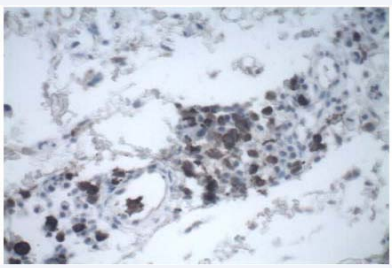

Figure 1 Rash on the back. The color of the rash was unusual and diffused.

The bone marrow smear to active bone marrow hyperplasia, M/E, was 1.47/1. 2.5% immature plasma cells .Biopsy and flow cytometry of the bone marrow was completely normal, including a normal kappa-lambda ratio We also used monoclonal antibodies against surface immunoglobulin kappa and lambda, showing normal expression of both. Serum protein and immunofixation electrophoresis demonstrated an elevated gamma globulin fraction (54.8%;reference range,9%-16%), with a diffuse polyclonal pattern and no monoclonal band. Bence-Jones protein was not detected in our studies. X-ray of the skull and upper jaw, mandible, and cervical bone showed destruction (Figures 2). This destruction wasn't an osteolytic lesion, causing us to realize the misdiagnosis of MM.

The initial skin biopsy specimen obtained from abdominal skin demonstrated hyperkeratosis of the skin, acanthosis, epidermal melanocytes that had increased dermal vascular peripheral plasma cells as the primary key in the shallow infiltration of the group, and no heteromorphism (Figures 3). Immunohistochemical stains showed CD38/CD138 positive, and CD56 negative cells. The kappa and lambda immunoglobulin light chains demonstrated a 2:1 ratio, which confirmed the polyclonality of plasma cells. Other test results are shown in (Table 1).

In summary, the patient was treated with thalidomide and dexamethasone for 6 cycles after her initial diagnosis of MM. She reported no obvious changes, and laboratory examinations showed no notable changes, so the patient elected to stop treatment. About 6 months after discontinuing her treatment, and a year after her diagnosis of MM, the patient came to see me. She received serum protein and immunofixation electrophoresis, a bone marrow biopsy, and a bone review. No changes were found. After a 9-month review, skin biopsies again supported the diagnosis of CSP. On follow-up at 50 months, the patient was in good condition with no obvious discomfort.

Table 1: Laboratory data.

Variable	Reference range (adults)	Patient data on admission
White cell count ($\times 10^9$/L)	4.0-10.0	1.3
Hemoglobin (g/L)	11-16	7.0
Platelets ($\times 10^9$/L)	100-300	230
Erythrocyte sedimentation rate (mm/h)	0-15	140
C-reactive protein (mg/L)	<10	86
Beta-2-microglobulin (mg/L)	<2.5	3.84
Serum albumin (g/L)	40-55	21.8
Serum globulin (g/L)	20-30	71
Lactate dehydrogenase (U/L)	135-215	68
Interleukin-6 (ng/L)	108.85 ± 41.48	235
Serum calcium (mmol/L)	2.25-2.75	1.91

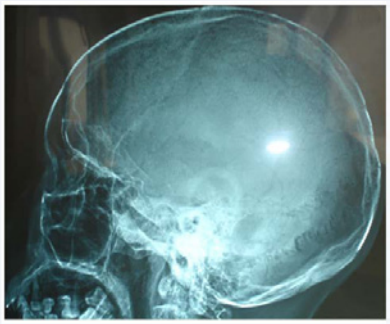

Figure 2 X-ray of the skull. X-ray shows discontinuity of bone plates and bone destruction.

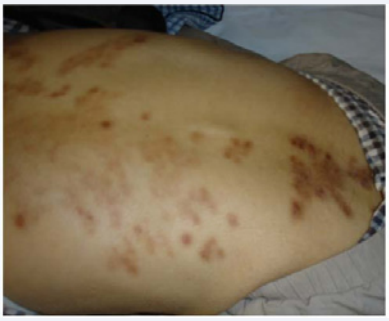

Figure 3 Pathological features of the skin. Infiltrates of mononuclear cells with numerous plasma cells in the derma (immunohistochemical staining showed CD138 positive. Diaminobenzidine staining, ×100 magnification).

DISCUSSION

CSP is a rare disease characterized by infiltration of the skin by polyclonal plasma cells associated with polyclonal gammopathy and variable systemic manifestations. Clinically, it is characterized by multiple red-brown plaques and flat tumors, mainly on the trunk. Histologic findings show a dermal infiltrate composed predominantly of polyclonal plasma cells [5,6]. In all patients, examination of a skin biopsy specimen has revealed the presence of a dermal nodular and perivascular mixed-cell infiltrate with a predominance of plasma cells. The epidermis was not involved in this case. The plasma cells did not reveal any atypical features, and immunohistological staining showed that the plasma cells were positive for CD138. Expression of kappa and lambda immunoglobulin light chains revealed polyclonal expression of both immunoglobulin light chains. Immunoglobulin heavy chain gene rearrangement was performed according to standard procedures.

To the best of our knowledge, this is the first reported case of CSP with bone pain. The patient had been misdiagnosed as having multiple myeloma in a local hospital. Although bone x-ray findings in patients with abnormal bone lesions are not specific, they should be used in conjunction with other forms of disease identification including monoclonal protein studies of bone marrow. According the diagnostic criteria our findings does not support the diagnosis of MM. The diagnosis of CSP is mainly dependent on skin pathology, as the skin biopsy of this patient was essential for a proper diagnosis.

Multiple myeloma is a malignant tumor and CSP is a benign disease; therefore, the ability to distinguish between the two is important to patients.

CONSENT

Written informed consent for publication of data was obtained from the patient.

COMPETING INTEREST

No potential conflict of interest relevant to this article was reported. We have no personal or financial conflicts of interest associated with the preparation and publication of this manuscript.

AUTHORS' CONTRIBUTIONS

The authors declare that there were no conflicts of interest in this work. Yuping Zhong designed and performed the study. Jiajia Zhang collected the data. Yuping Zhong wrote the paper.

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