PRIMARY PLASMA CELL LEUKEMIA
(A COMPREHENSIVE ANALYSIS OF 44 CASES)

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Four cases of primary plasma cell leukemia (PPCL) admitted to Zhongshan Hospital from 1959 to 1987 are reported with a review on additional 40 cases reported in China. Comparing with the 57 cases of multiple myeloma (MM) treated in our hospital, the following features were observed in PPCL: (1) The age was younger, with a mean of 45.2 years, 34.1% of the patients were under 40 years. (2) Onset was abrupt. Duration from onset to diagnosis was 2 months or less in 77% patients but never beyond 6 months. (3) 81.8% patients had liver enlargement, 59.1% splenomegaly and 61.4% sternum tenderness. (4) All patients showed marked anemia with an average hemoglobin of 65 g/L. BPC count was less than $10^9$/L in 76% patients and WBC was more than $10^9$/L in 77%. (5) Plasma cell number in the marrow was markedly increased with an average of 69%, of which the blast cells and immature forms were predominant. (6) No destruction of bones was shown on X-ray film in 68.3% patients. (7) The response to chemotherapy was poor with a total response rate of 18% and a mean survival of 2 months. All the above-mentioned clinical features were significantly different from those of MM. In addition, these two diseases were also different in cytology, cytogenetics and ultrastructure. Therefore, PPCL should be considered as a special type of acute leukemia distinct from MM. High dose of alkylating agents in combination with autologous bone marrow transplantation might improve the prognosis.

Primary plasma cell leukemia (PPCL) was rare. There were different views in regard to the relationship between PPCL and multiple myeloma (MM). In this paper, the authors reported four cases of PPCL admitted to our hospital from 1959 to 1987 inclusive with a review of additional 40 cases reported in Chinese medical literature. The difference in diagnosis and treatment of PPCL and MM was discussed.

MATERIALS AND METHODS

Diagnostic Criteria

The criteria of the diagnosis of plasma cell leukemia (PCL) was adopted as proposed by Kyle. In PPCL, the diagnosis was made when the patient had plasma cell in the peripheral blood above 20% at the onset of illness, with the absolute plasma cell level above $2.0 \times 10^9$/L. In most cases, the patient might have fever, bony ache, marked anemia and hemorrhage. In physical examination, hepatosplenomegaly, lymphadenopathy and sternum tenderness were noticed. However, the patient had no previous history of myeloma. In secondary plasma cell leukemia (SPCL) patient might show almost similar clinical feature, but had previous history of MM, macroglobulinemia, lymphoma, chronic lymphocytic leukemia, and amyloidosis etc., plasma cell in the peripheral blood increased markedly only at the later stage of the disease.

Clinical Data of PPCL

Incidence: Among seven patients with PCL in our hospital, only four were PPCL (Table 1). A total of 55 patients of PCL was recorded in the reports from 32 journals in China, among them, 44 were PPCL and 18 SPCL. For the latter, 16 patients were secondary to MM, 2 had aplastic anemia at the onset.

Age and Sex: Age range of the patients with PPCL in total 44 cases was from 9 months to 77 years, median 45.2 years, with 34.1% under age of 40. There were 33 male and 11 female, ratio 3:1.

Clinical Manifestations: The onset of disease was abrupt. In 20% of patients, the time required for establishing diagnosis was 2 weeks or less and in 77.1% within 2 months, but none beyond 6...
Table 1. Clinical data of 4 patients with PPCL

<table>
<thead>
<tr>
<th>Case symptoms</th>
<th>physical signs</th>
<th>blood count</th>
<th>% PC in BM</th>
<th>X-ray of bone</th>
<th>R</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fever, hemoptysis for 2 months</td>
<td>3.0</td>
<td>+</td>
<td>65.0</td>
<td>NP (NR)</td>
<td></td>
<td>4mo</td>
</tr>
<tr>
<td>Fever, lumbago, gingival bleeding for 40 days</td>
<td>2.0</td>
<td>-</td>
<td>70.0</td>
<td>Osteoporosis (NR)</td>
<td></td>
<td>40d</td>
</tr>
<tr>
<td>High fever, cough, hemoptysis for 2 months</td>
<td>2.0</td>
<td>-</td>
<td>75.0</td>
<td>H (NR)</td>
<td></td>
<td>1mo</td>
</tr>
<tr>
<td>Fatigue, anorexia, hematuria for 2 months</td>
<td>1.5</td>
<td>-</td>
<td>85.0</td>
<td>COCP (NR)</td>
<td></td>
<td>21d</td>
</tr>
</tbody>
</table>


Laboratory Findings: For 44 patients of PPCL, hematological studies showed Hb 27—140 g/L with a mean value of 65.3 g/L, WBC 3.5—118×10⁹/L with a mean 25.2×10⁹/L, platelet 2.0—220×10⁹/L with a mean 76.2×10⁹/L at the onset of illness. Plama cell ranged from 14—96% in the peripheral blood (mean value 47.6%), with more than 20% in 41 patients. Absolute plasma cell count ranged from 0.63—82.7×10⁹/L (mean value 11.58×10⁹/L), above 2×10⁹/L in 40 patients. Bone marrow examination showed hypercellularity in 57.5%, hypocellularity in 2.5%, and normocellularity in the others. The number of plasma cell in the marrow was 29.5—94% with an average of 69.1%, predominantly blast cell and immature forms (average 54.8%), mature plasma cell were 14.3%. Serum immunoglobulin determined in 20 patients showed increased monoclonal IgG in nine, monoclonal IgM in one, and monoclonal IgA in one, 8 patients had normal Ig and decreased Ig in one patient. 11 of 37 patients showed Bence-Jones proteinuria. The study of kidney function showed...