

Case Report

PRIMARY PLASMA CELL LEUKEMIA- A CASE REPORT

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ABSTRACT

Plasma cell leukemia is a rare disease entity where the number of clonal plasma cells in the peripheral blood exceeds $2 \times 10^9/l$ or 20% of the total leukocyte count. This plasma cell leukemia can be primary which is arising de novo or secondary as in the course of plasma cell myeloma. A 40 year old female who came to the outpatient department with complaints of low back pain for 3 months and had hepatosplenomegaly. Patient was investigated for the same and the peripheral smear showed 20% plasmacytoid cells. Multiple lytic lesions were noted in x ray skull. Bone marrow aspirate and biopsy revealed a hyper cellular marrow. Marrow showed chiefly plasma cells (56%) with many binucleate, trinucleate forms and multinucleate forms resembling tumour giant cells were noted. Marrow elements were decreased. Chemotherapy was initiated and the patient had successfully completed four cycles. This case is presented because of its rarity, presenting at a younger age, primary type of plasma cell leukemia, presence of organomegaly and unique morphology of the cells in the blood and bone marrow.

Key Words: *Plasma Cell Leukemia, Plasmacytoid Cells, Primary, Lytic Lesions*

INTRODUCTION

Plasma cell leukemia is a rare disease entity where the number of clonal plasma cells in the peripheral blood exceeds $2 \times 10^9/l$ or 20% of the total leukocyte count(McKenna 2008). This plasma cell leukemia can be primary which is arising de novo or secondary as in the course of plasma cell myeloma(Gertz 2010). These neoplastic plasma cells which are usually found in the blood and bone marrow can also be seen in extramedullary sites like liver, spleen, pleural effusion, ascitic fluid and cerebrospinal fluid(McKenna 2008).

CASES

A 40 year old female came to the outpatient department with complaints of low back pain for 3 months and history of fever for 17 days. On examination patient had hepatosplenomegaly. Patient was investigated with complete blood count and peripheral smear. Her results are as follows Hb 7.1g/dl, WBC total count $7.2 \times 10^9/l$ Differential count – neutrophil 20%, lymphocyte 55%, monocyte 2% eosinophil 3%, and plasma cells 20%. Platelet count $0.54 \times 10^9/l$. Peripheral smear showed 20% plasmacytoid cells which were large cells with central nucleus and moderate amount of basophilic cytoplasm. Few of the cells had prominent nucleoli. X ray skull showed multiple lytic lesions. With the history of low back pain, lytic lesions in the skull and 20% plasmacytoid cells in the peripheral smear a diagnosis of plasma cell leukemia was made. Bone marrow aspirate and biopsy showed a hyper cellular marrow. Marrow showed chiefly plasma cells (56%). Many binucleate, trinucleate forms of plasma cells with many multinucleate forms resembling tumour giant cells were noted. Erythroid series, myeloid series and megakaryocytes were decreased. Chemotherapy was initiated and the patient had successfully completed four cycles.

DISCUSSION

Primary plasma cell leukemia is important to recognize because of its rarity of occurrence and unusual presentation. There are two types of plasma cell leukemia, one is primary and the other is secondary. In a study conducted by the SEER group between 1973 and 2004 approximately 49,000 patients with myeloma were identified. This study did not distinguish primary and secondary forms of plasma cell leukemia. The incidence was found to be 4 cases per 10,000,000 persons per year in Europe. Secondary

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PCL occurs as a progression of the disease in 1 to 4% of all cases of myeloma. The reported incidence of PCL ranges from 1.6% to 5% in different series. The incidence of primary PCL is very rare and reported to occur in less than one in a million (Naseem 2012)

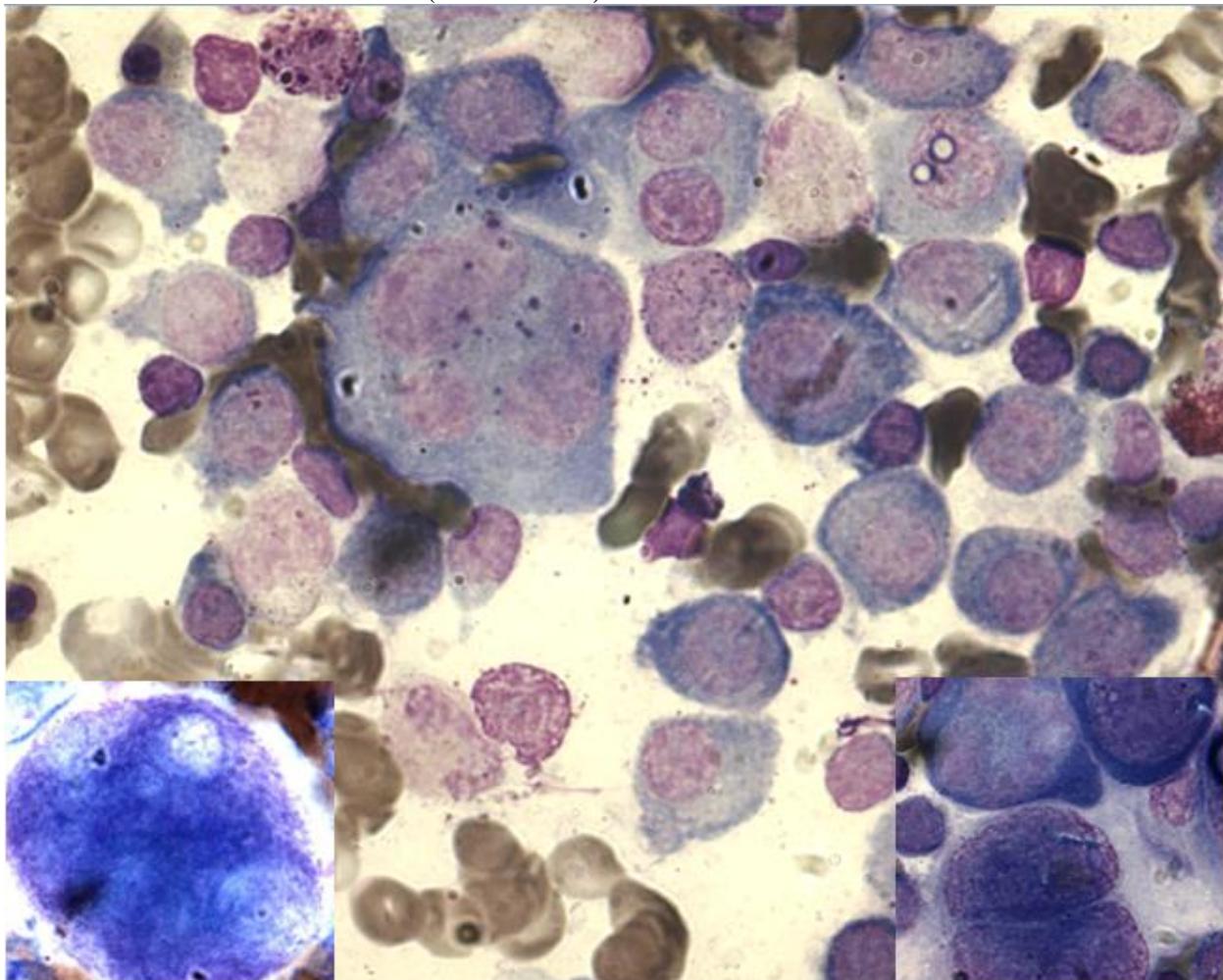


Figure 1: Shows plasma cell with binucleate, multinucleate forms, inset shows pleomorphic plasma cells.

Plasma cell leukemia becomes important because of its rarity of presentation, clinical presentation, morphological difference from myeloma cells, and expression of immune markers and survival of the patient.

This case is presented because of its rarity, presenting at a younger age, primary type of plasma cell leukemia, presence of organomegaly and the morphology of the cells in the blood and bone marrow. The low power view of this smear appeared like metastasis. Examination under oil immersion revealed the presence of multinucleated plasma cells. A similar case of 51 year old male patient with history of fever and altered sensorium, with peripheral blood plasma cells of 24% was reported by Abid Jameel (Jameel 2005). Another case report by Singh *et al.*, (2009) was reported in a 70 year old male who presented with weakness and cervical lymphnodes. This case showed plasma cells and plasmablasts in the bone marrow. The present case in our study showed multinucleate, giant plasma cells in the bone marrow. In both the cases the prognosis was bad with death of the patient after diagnosis in few months. A similar case of pleomorphic morphology of plasma cell in plasma cell leukemia was reported by John *et al.*, (2007). The present case in our study is comparatively doing well with completion of 4 cycles of chemotherapy at the

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end of 8 months. The chemotherapy protocol was Thalidomide with steroids. The plasma cells in the peripheral smear have reduced to zero. In course of few months she developed steroid induced hyperglycemia. She was treated for the same.

Summary

Primary plasma cell leukemia is very rare. The incidence of plasma cell myeloma ranges between 2 and 4% of all myelomas. About 60% of plasma cell leukemia is primary and rests 40% are secondary, as a part of terminal phase in the course of plasma cell myeloma. Clinically the age of presentation is younger with presence of hepatosplenomegaly and fewer lytic lesions in the bone when compared to the plasma cell myeloma. The morphology of the plasma cells in this case was highly pleomorphic with many multinucleated ones which resemble like the tumor giant cells. Although the prognosis was bad this patient had completed 4 cycles of chemotherapy.

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