

Case Report

Primary Plasma Cell Leukemia- A Rare Case Report

S.R. Mehdi¹, Kshama Tiwari², Amrisha Jaiswal³, Rasha Zia Usmani³, Tanya Tripathi³

¹Professor & HOD, Department of Pathology, Era's Lucknow Medical College, Lucknow

²Assistant Professor, Department of Pathology, Era's Lucknow Medical College, Lucknow

³Junior Resident, Department of Pathology, Era's Lucknow Medical College, Lucknow

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 denovo or secondary as in the course of plasma cell myeloma. A 48 years old male patient came to the emergency outpatient department with complaints of acute renal failure without any prior history of diabetes or hypertension. On examination patient also had hepatosplenomegaly. Peripheral smear showed 56% plasmacytoid cells. Bone marrow aspirate and biopsy revealed a hyper cellular marrow. Marrow showed chiefly plasmablasts (32%) with plenty of abnormal plasmacytoid cells (21%). Other marrow elements were grossly reduced. 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.

Keywords: Plasma cells, plasmacytoid lymphocytes, primary plasma cell leukemia, multiple myeloma

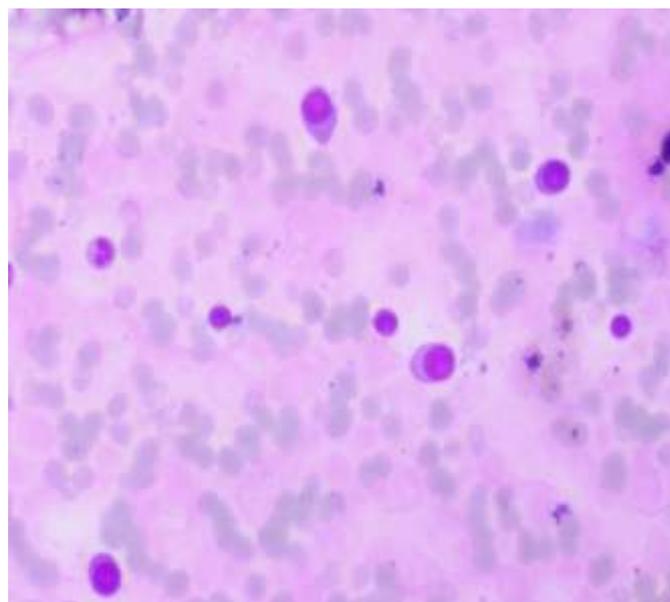
INTRODUCTION

Plasma cell leukemia (PCL) is a rare disorder. Patients may either present de novo (primary PCL), or PCL may occur during the course of multiple myeloma (secondary PCL).¹ In a study conducted by the SEER group between 1973 and 2004, the incidence of plasma cell leukemia was found to be 4 cases per 10,000,000 persons per year in Europe. Secondary PCL occurs as a progression of the disease in 1 to 4% of all cases of myeloma. The incidence of primary PCL is very rare and reported to occur in less than one in a million.² Plasma cell leukemia is diagnosed when the number of clonal plasma cells in the peripheral blood exceeds $2 \times 10^9/l$ or 20% of the total leukocyte count.³ These neoplastic plasma cells can also be seen in extra-medullary sites like liver, spleen, pleural effusion, etc.³ According to a survey, PPCL may account for about 60% and secondary PCL for about 40% of cases.⁴ The average age of diagnosis of primary PCL is 55 years old, slightly younger than in myeloma. Average age of patients diagnosed with secondary PCL is 66 years old.⁵

CASE REPORT

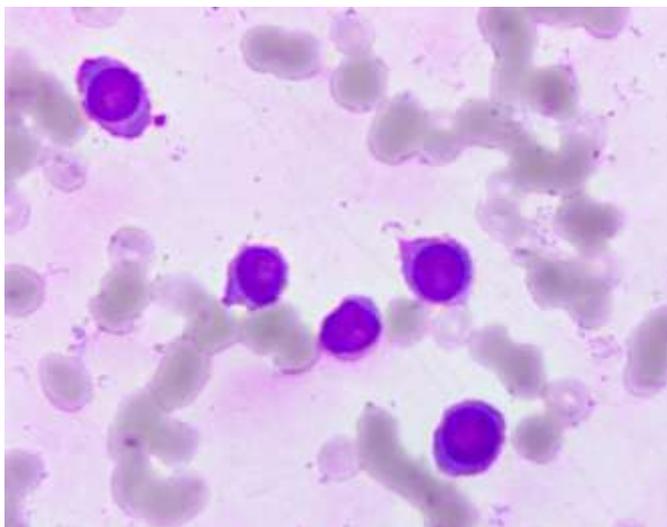
A 48 yrs old male patient came in emergency OPD with the chief complain of acute renal failure with a serum urea 194.6 mg/dl and serum creatinine of 8.5 mg/ . His previous medical and family histories were unremarkable. On examination there was moderate hepato-splenomegaly and the blood sample was send to pathology department for complete blood count and general blood picture. His results were as follows -Hb 4.5 g/dl, total leucocyte count 17000 cells/ cu mm with a differential of polymorphs 42 %, monocytes 01%, eosinophil 01% and

plasma cells with plasmacytoid lymphocytes 56% . Platelets were grossly reduced with a count of 50,000/ cu mm. On bone marrow examination, aspirate showed a hypercellular marrow with predominant plasmacytoid cell population hyperplasia consisting chiefly of plasmablasts (32%) with plenty of abnormal plasmacytoid cells (21%).



Peripheral blood smear showing plasma cells (high power)

All other series like erythroid , myeloid and megakaryocyte were grossly reduced by smear.



Peripheral blood smear showing plasma cells (Oil immersion)

With this haematological findings, diagnosis of Plasma cell leukemia was made and patient further evaluated for S. Ca^{++} , S. LDH and β_2 microglobulin. The lab reports were as follows- S. Ca^{++} - 10.5 mg%, S. LDH -724 mg/dl, and β_2 microglobulin-8.45; showing though normal S. Ca^{++} , but markedly elevated S. LDH and β_2 microglobulin values thus confirming the diagnosis of plasma cell leukaemia. To rule out presence of underlying plasma cell dyscrasias, full skeletal X-Ray was recommended, which showed no lytic lesions anywhere in the body; excluding the possibility of pre-existing multiple myeloma.

DISCUSSION

Plasma cell leukemia is important to recognize because of its rarity of occurrence and unusual presentation. There are two types of PCL: Primary PCL – diagnosed in patients with no history of myeloma, Secondary PCL – occurs after a previous myeloma diagnosis and arises when myeloma progresses to PCL. The likelihood of myeloma patients developing secondary PCL is very low (about 0.5 - 1%). Secondary PCL is thought to be caused by an accumulation of specific genetic changes that occur within myeloma cells as the myeloma progresses in certain patients. It is estimated that 1 per million of the general population are diagnosed with primary PCL each year, while approximately 1 in 100 myeloma patients will go on to develop secondary PCL.

This case is presented because of its rarity, presenting at a younger age, primary type of plasma cell leukemia, presence of organomegaly, morphology of the cells in the blood and bone marrow and poor prognosis of the patient.

A 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.⁶ Another case report by Singh et al., was reported in a 70 year old male who presented with weakness and cervical lymphnodes.⁷ This case showed plasma cells and plasmablasts in the bone marrow. The present case in our study also showed plasmablasts in the bone marrow. In both the cases the prognosis was bad with death of the patient after diagnosis in few months. Prognosis in our case was also

not good as the patient died soon after the chemotherapy has been started. A similar case of pleomorphic morphology of plasma cell in plasma cell leukaemia was reported by John et al.⁸ Thus a very few case reports are present related to plasma cell leukaemia documenting its rare incidence.

SUMMARY

Primary 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.

We have studied and reported this case of primary plasma cell leukemia, because age of presentation of patient was only 48yrs, quiet younger than what is the documented age i.e. 55yrs, presence of organomegaly in the form of hepatosplenomegaly, absence of lytic lesions of bone and most importantly very poor prognosis of the disease.

REFERENCES

- [1] Gertz MA, Buadi FK. (2010). Plasma cell leukaemia. *Haematologica* 95(5) 705-707.
- [2] Naseem S, Kaur S, Gupta R, Kashyap R, Nitayand S (2012). Plasma cell leukaemia; Case series from a tertiary centre with review of literature. *Indian Journal of Hamatology and Blood Transfusion* 28(1):10-14.
- [3] McKenna RW, Kyle RA, Kuehl WM, Grogan TM, Harris NL, Coupland RW (2008). Plasma cell neoplasms. In: *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*. IARC Lyon. 200-213.
- [4] Noel P, Kyle RA. Plasma cell leukemia: an evaluation of response to therapy. *Am J Med* 1987;83: 1062–1068.
- [5] Plasma cell leukaemia [Internet]. Edinburgh: Myeloma UK; Aug 2008[updated feb 2017]. Available from: <https://www.myeloma.org.uk/Myeloma-UK-Plasma-Cell-Leukaemia-Infosheet.pdf>
- [6] Jameel A(2005). Plasma cell leukaemia: Case report of a rare and aggressive variant of multiple myeloma *Journal of Pakistan Medical Association* 55(10) 452-453
- [7] Singh T, Premalatha CS, Sanjeevan KV et al., (2009). Ig A Plasma cell leukaemia. *Journal of Lab Physicians* 1(1) 19-21.
- [8] John S, Feroze M, Supriya NK, Aisabi KA (2007). *Indian Journal of Pathology Microbiology*. 50(1) 45.