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PRIMARY PLASMA CELL LEUKEMIA- A CASE REPORT

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ABSTRACT

Plasma cell leukemia is a rare variant of multiple myeloma and this term is applied when the absolute plasma cell count is greater than $2x10^9$ /L or the relative plasma cell number is greater than 20% of the peripheral white blood cells. PCL may be primary or secondary. A 55 year old male presented to our outpatient department with complaints of lower back pain for four months, low grade fever off and on for two months, generalised weakness and loss of appetite for 20 days without prior history of multiple myeloma. His peripheral blood smear showed 26% plasmacytoid cells and bone marrow revealed predominantly plasma cell population (80%) with binucleate and trinucleate forms. Chemotherapy was started but unfortunately patient had expired. This case is presented due to its rare occurrence, presence of organomegaly and poor outcome.

Key words: Plasma cell leukemia

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INTRODUCTION

Plasma cell leukemia is a rare variant of multiple myeloma and this term is applied when the absolute plasma cell count is greater than $2x10^9$/L or the relative plasma cell number is greater than 20% of the peripheral white blood cells and PCL may be primary or de novo, when the patient is diagnosed first time without prior history of myeloma or secondary PCL, when a patient with previously diagnosed myeloma undergoes leukemic transformation.[1] PCL is found in 2 to 5 % of cases of myeloma and plasma cells may be found in extramedullary sites such as spleen, liver, pleural fluid,ascites and cerebrospinal fluid.[1] Hepatosplenomegaly , lymphadenopathy, and lytic bone lesions are more common in primary PCL and secondary PCL respectively.[2] The age range of patients with PCL is 50- 60 years with approximately equal proportion of male and female patients.[3]

CASE REPORT

A 55 year old male presented to outpatient department of Lala Lajpat Rai hospital, Kanpur during the month of May 2015 with complaints of lower back pain for four months, low grade fever off and on for two months, generalised weakness and loss of appetite for 20 days. There was no prior history of multiple myeloma or other severe illness. On examination he had mild hepatosplenomegaly. This biochemistry investigation revealed random blood sugar level of 120 mg/dl, kidney function tests were abnormal with serum urea 90mg/dl and serum creatinine was 10.8 mg/dl. His complete blood count showed Hb- 5.4 gm/dl, WBC- 19,400cells/cumm, platelets -75,000/cumm, and his ESR was 120 mm/hr. Leishmen stained peripheral blood smear showed normocytic normochromic RBCs with increased rouleaux formation, mildly raised TLC and differential count showed 26% plasmacytoid cells with moderate amount of basophilic cytoplasm

and eccentric nuclei and neutrophils - 55% ,lymphocytes -13%, monocytes - 4%, eosinophils-2% and platelets were reduced in number. This was an incidental finding on smear and was reported as plasma cell dyscrasia and advised for urine examination, serum protein electrophoresis and other specific tests for confirmation. Serum protein electrophoresis revealed presence of thick M band. Total protein was 10.10 gm/dl, fraction of albumin -3.57 g/dl ,α1-0.86g/dl, α2-1.26g/dl, β-0.85g/dl and γ -3.57g/dl and A/G ratio was 0.55%. Gamma fraction was markedly increased. Urine showed presence of Bence Jones protein and mild hepatosplenomegaly was present on USG.

Bone marrow aspiration was done from right posterior superior iliac spine, bone marrow aspirate smears diluted with sinus blood showed increased cellularity, predominantly plasma cells (80%). Many binucleate and trinucleate forms of plasma cells in varying stages of maturation were noted. Erythroid, myeloid and megakaryocytic series were suppressed. Patient was uncooperative so bone marrow biopsy was not done but diagnosis of primary PCL was established on the basis of above findings and patient was referred to higher centres where he was put on chemotherapy but unfortunately he had expired after one month of diagnosis.

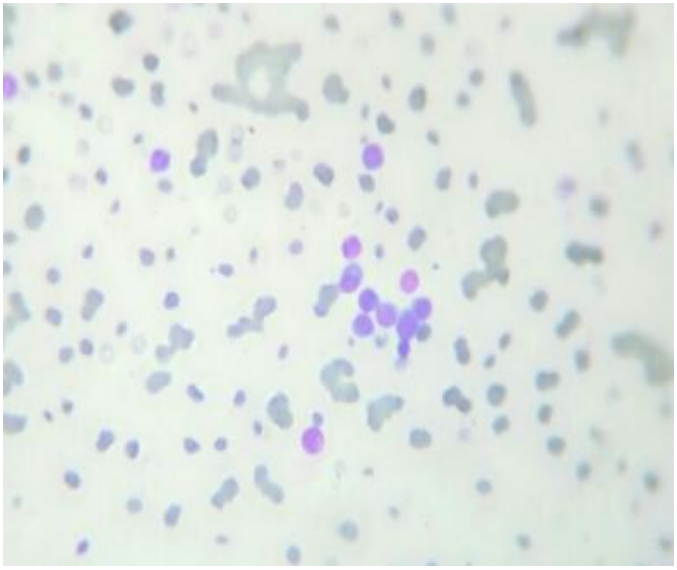

Figure-1 (4x10,40X, Leishman Peripheral blood smear-showing increased rouleaux formation along with aggregates of plasma cells

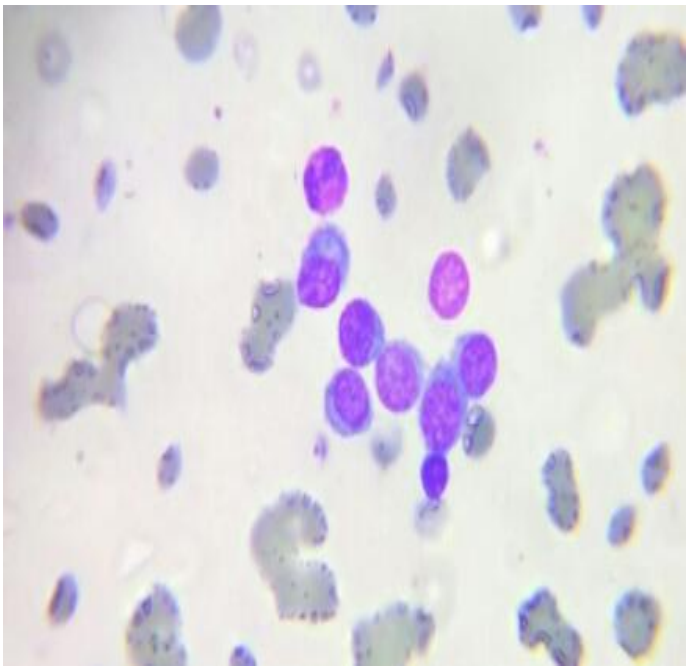

Figure-2 (40x10,400X,Leishman Peripheral blood smear-showing increased rouleaux formation along with aggregates of plasma cells

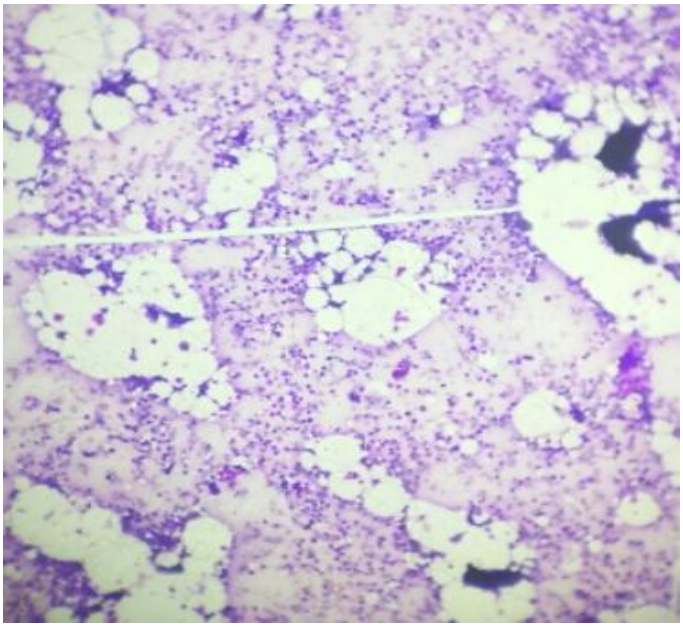

Figure-3 (4x10, 40X Giemsa – Bone marrow aspirate smear-showing small marrow particles and dilution with sinus blood

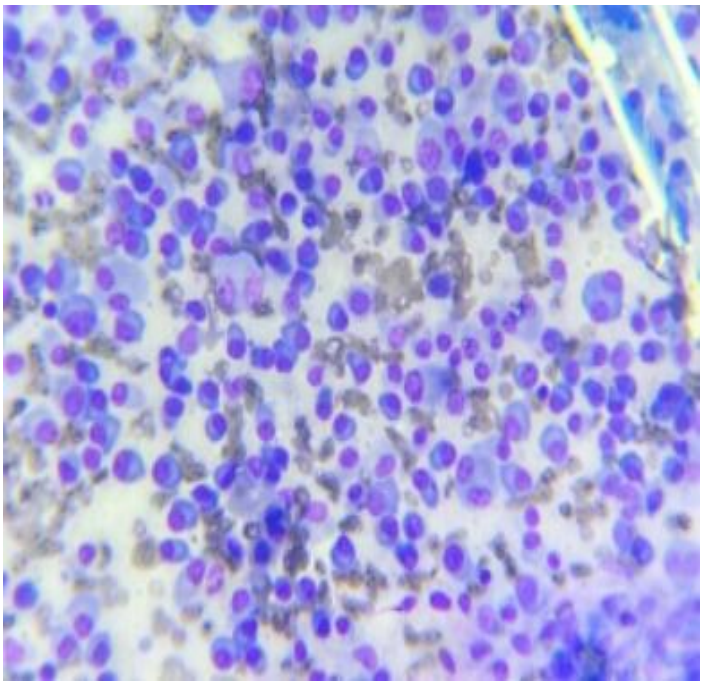

Figure-4 (40x10,400X Giemsa – Bone marrow aspirate smear-showing proliferation of Plasma cells replacing erythroid, myeloid and megakaryocytic series cells,binucleated forms of plasma cells

DISCUSSION

The exact incidence of primary plasma cell leukemia is unknown however in some studies the incidence of primary PCL has been reported to less than one in a million. [4] PCL occurs as a progression of a disease in 1 to 4 % of all cases of myeloma and reported incidence of PCL ranges from 1.6% to 5% in different series. [4] PCL is an important entity to recognise because of its rare occurrence and poor prognosis. .Both types of plasma cell leukemia have similar clinical presentation, resembles late stage multiple myeloma. Patients may present with anemia, pancytopenia, recurrent bacterial infections, and bleeding manifestations or with signs and symptoms of renal insufficiency**. [3]**

Patient may present with complaints of weakness, low backache, fever, hepatosplenomrgaly and cervical lymphadenopathy and > 20 % plasma cells in peripheral blood and in bone marrow**. [5, 6]** Pleomorphic morphology of plasma cells is also reportd**. [7]** The present case presented with lower back pain, low grade fever, generalised weakness and mild hepatosplenomegaly and showed 26% plasma cells in PBS and 80% plasma cells in bone marrow with binucleated and trinucleated morphology. Fonseca et al reported the case of primary PCL with no M band and raised α2 globulin fraction on protein electrophoresis whereas in our case there was a thick monoclonal M band with massively raised gammaglobulin fraction.**[3]** Plasma cells in primary PCL frequently display a more immature phenotype expression of pan B cell antigen and CD20 has been shown in 50% of PCL cases as compared to 17% of multiple myeloma cases. An increased incidence of cytogenetic abnormalities has been reported in PCL as compared to multiple myeloma**.[3,8]** Response to treatment of PCL is very poor and optimal regimen for primary PCL is not firmly established however intensive multi agent chemotherapy and bone marrow transplant should be considered especially in younger patients.**[9]** Patients with primary PCL may initially respond better to chemotherapy including single agent drugs commonly used in multiple myeloma whereas resistant disease is expected and a median survival of less than six months for both types of PCL has been observed**.[3]** In the present case prognosis was bad with death of the patient after the diagnosis in few months.

CONCLUSION

Primary PCL is an important variant of multiple myeloma with aggressive nature and rarity of occurrence and has poor prognosis. This case is reported due to its unusual presentationwith hepatosplenomegaly.

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