

AN UNUSUAL PRESENTATION OF PRIMARY PLASMA CELL LEUKEMIA AS A PSYCHIATRIC DISORDER: A DIAGNOSTIC DILEMMA- A CASE REPORT AND REVIEW OF LITERATURE**Anshu Gupta¹, Chhavi Gupta², Anju Kapoor³**

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ABSTRACT

Plasma cell leukemia (PCL) is a rare disorder that can develop spontaneously (primary) or evolve in patients with multiple myeloma (secondary). Histogenetically, plasma cell leukemia is derived from terminally differentiated B cells. It is diagnosed by the presence of absolute plasma cell count >2000/cumm or >20% circulating plasma cells in blood. We report a rare case of primary plasma cell leukemia in patient with unique presentation of a psychiatric illness having hallucinations, mood changes and loss of insight. The patient had past history of pulmonary tuberculosis and developed de novo PCL as there was no previous history of multiple myeloma. Clinical spectrum, cytomorphological features and prognosis of PCL are also discussed in this case report.

Key words: Psychiatric, Primary, Plasma cell, Leukemia, Multiple myeloma

INTRODUCTION

Plasma cell leukemia (PCL) constitutes 2-4% of plasma cell dyscrasias. It is rare and clinically aggressive variant of plasma cell myeloma^[1]. It is defined as >20% plasma cells in peripheral blood or an absolute plasma cell count of >2000/cumm^[2]. Two types of PCL are recognized -Primary and Secondary. Primary PCL is a de novo disease with no previous evidence of multiple myeloma^[3]. The clinical presentation of PCL as a psychiatric disorder is rare and which can be misleading causing diagnostic dilemma if not work up properly. Psychiatric symptoms secondary to PCL typically present after the development of hypercalcemia and uremia^[4].

CASE STUDY

70 yrs old female patient brought to our hospital with psychiatric complaints such as hallucinations, depressed mood and self-care failure for three months. She also had auditory and visual hallucinations. All the routine investigations were normal. She was then diagnosed as bipolar affective disorder & was given antipsychotic drugs. One month later, patient was again brought to the hospital with similar complaints but of increased severity. An additional

antipsychotic drug was included in the previous regimen. All the routine investigations were within normal range this time also. Five weeks later, the patient landed in Emergency Department with neurological complaints of weakness of lower limbs, generalized myalgia, fatigability and back pain over dorso-lumbar region. The patient was admitted. During hospital stay, she had developed loss of bowel and bladder sensation along with incontinence. She had past history of pulmonary tuberculosis also and took anti tubercular treatment one year ago.

On clinical examination, bony tenderness was present over dorso- lumbar region. There was no evidence of hepatosplenomegaly or lymphadenopathy on per abdomen examination. Routine Biochemistry investigations revealed random blood sugar level- 60mg/dl. Kidney function tests were deranged with serum urea 141mg/dl, serum creatinine 2.0mg/dl and serum uric acid 7.0mg/dl. Total serum calcium was 11.8mg/dl. Hematological investigations showed hemoglobin (Hb) 9.0gm%, ESR 92mm in 1st hour. Total leukocyte count (TLC) - 16700 cells/cumm with neutrophils 68%, lymphocytes 07%, eosinophils 01% , monocytes 01% and 23% plasma cells. Absolute plasma cell count was

3841 /cumm. Microscopically, Leishman stained peripheral blood smear revealed increased plasma cells with binucleated forms and mott cells (Fig.1). Platelet counts were within normal limits. This was an

incidental finding on smear and was reported as plasma cell dyscrasias. Further detailed work- up of the patient was done in this direction and specific tests were done for confirmation.

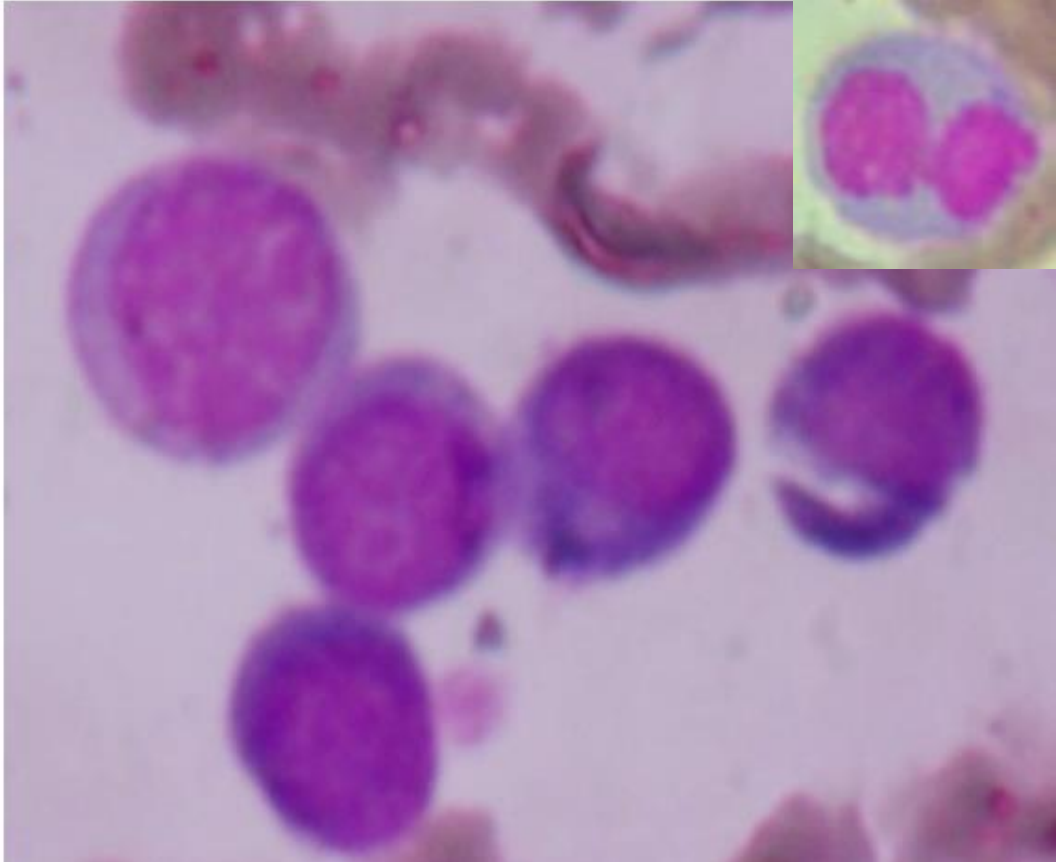


Fig.1. Photomicrograph of peripheral blood smear showing mature and immature plasma cells with binucleated form (inset). (Leishman x 100x)

On radiological examination, X-rays spine showed generalized osteoporosis and lytic destruction involving D8 and D9 vertebral bodies. MRI of dorso- lumbar spine revealed osseous destruction with altered marrow signal intensity involving D8 and D9 vertebral bodies with compression of the dorsal spinal cord. Based on overall findings, a diagnosis of primary plasma cell leukemia was made.

Serum electrophoresis revealed hypogammaglobulinemia and presence of M band of IgG type. Test for urinary Bence – Jones proteins was negative. Bone marrow aspirate showed fatty aspirate. Then, bone

marrow biopsy was taken in piece meal and examined microscopically. The biopsy showed hypercellularity with sheets of plasma cells which constituted 60% of nucleated cells of the marrow. Both mature and immature plasma cells seen .Immature plasma cells constitute 15% of total plasma cells. Immature plasma cells have eccentric to central nucleus with diffuse chromatin and inconspicuous nucleolus. There was paucity of erythropoiesis and myelopoiesis (Fig.2). On immunochemistry, the plasma cell showed strong CD38 positivity (Fig.3). Patient was put on multidrug chemotherapy & was advised close follow-up.

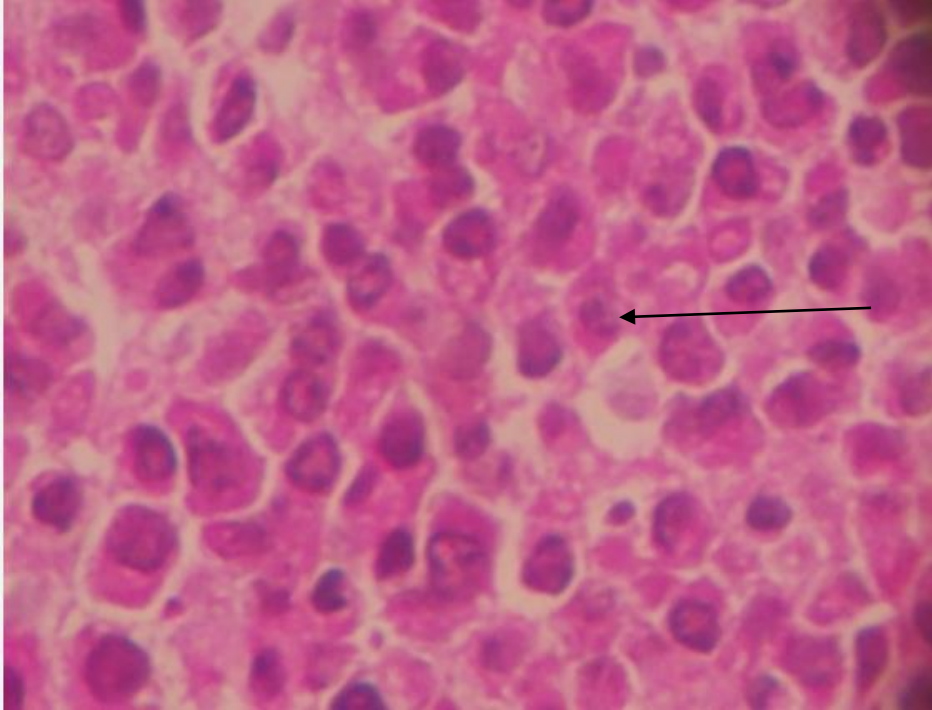


Fig2. Photomicrograph of bone marrow aspirate smear revealing sheets of plasma cells with atypical mitosis shown by arrow. Mature plasma cells have eccentric nucleus with perinuclear hof, dense chromatin and abundant eosinophilic cytoplasm (Papanicolaou x 40x).

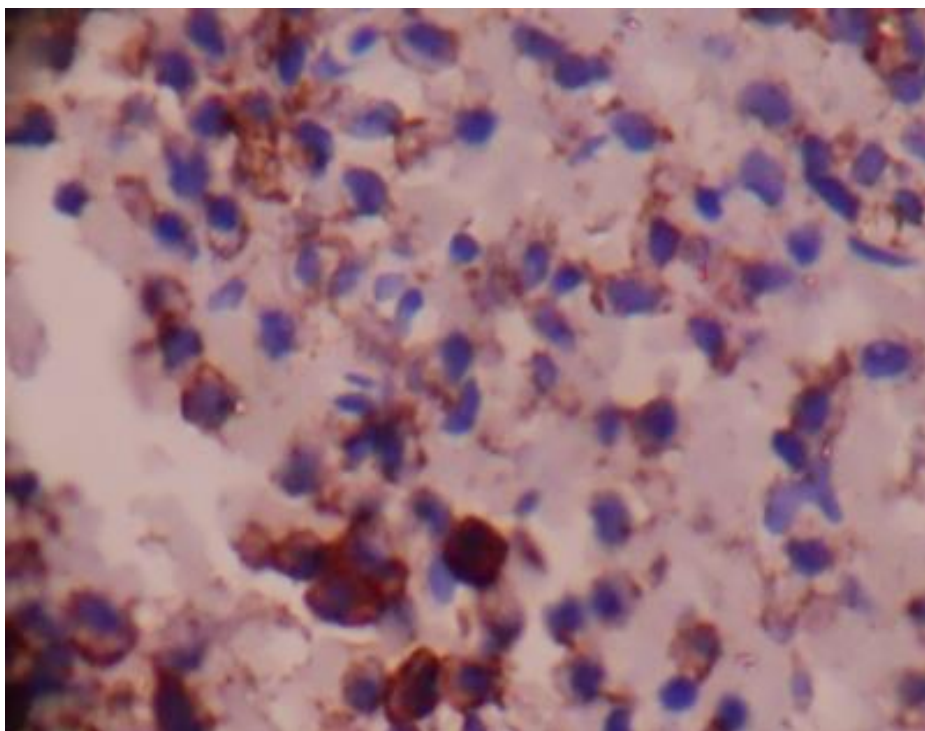


Fig.3. Photomicrograph of immunostaining for CD38 of bone marrow aspirate showing cytoplasmic positivity in mature and immature plasma cells. (Immunochemistry x 10x)

DISCUSSION

Two types of Plasma cell leukemia (PCL) are recognized -Primary and Secondary & can present as primary PCL in the absence of a previously diagnosed multiple myeloma (MM) or as secondary PCL, representing a leukemic

transformation of previously established MM [3]. Primary PCL constitute 60-70% of cases and the remaining 30-40% are secondary [5]. The major differences between primary and secondary PCL has been summarized in tabulated form on the basis of previous studies [6] and were given below:

	Primary	Secondary
Incidence	60-70%	30-40%
Age of presentation	5 th - 6 th decade	6 th decade
Onset	Abrupt	slow
Course of disease	Acute	Chronic
Hepatosplenomegaly	Present	Less common
Lymphadenopathy	Present	Less common
Survival time	Short	Short
Absolute plasma cell count	Higher	<Primary
Thrombocytopenia	Severe	moderate
Organ involvement	Extensive	Less extensive
Response to chemotherapy	Poor	Fair
Prognosis	Very Poor	Poor

Primary PCL is characterized by an aggressive clinical presentation. The clinical features of a psychiatric disorder in PCL are very rare and typically occur after development of hypercalcemia and uremia. Our case is unique because psychosis preceded the onset of PCL by up to three months without hypercalcemia and uremia [4]. Weiss et al found that one-third of myeloma patients were confused and suggested that a higher than expected incidence of delirium may be found in patients with multiple myeloma [7] Silberfarb et al also found that some multiple myeloma patients were delirious and others had depression [4].

Plasma cell dyscrasias are a common group of disorders having the proliferation of a single clone of immunoglobulin secreting cells as a common feature. [8] In the present case, the IgG type of monoclonal protein was evident on electrophoresis. Bone marrow is necessary for diagnosis of plasma cell leukemia, even if clinical, biochemical and radiological evidence is convincing. [9] The leukemic plasma cells have variable morphology. Our case showed marrow replacement by sheets of plasma cells which

constituted 60% of all nucleated cells. Bone marrow biopsy revealed both mature and immature plasma cells.

In PCL a higher expression of CD20 and CD 38 antigens is observed [10]. In our case; immunochemistry showed expression of CD38 which is a main plasma cell marker. Response to treatment of PCL is very poor and optimal regimen for primary PCL is not yet firmly established. However intensive multi agent chemotherapy and bone marrow transplant should be considered especially in younger patients [3]. This study gives an opportunity to discuss the clinical spectrum, cytomorphology and prognosis of this interesting condition. Consulting psychiatrists should be aware of the common occurrence of impaired cognition in patient with myeloma.

CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest. This manuscript has been read and approved by all the authors, that the requirements for authorship have been met, and that each author believes that the manuscript represents honest work.

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