

## Primary Plasma cell Leukemia- a Case Report

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### Abstract

Plasma cell leukemia is one of the most aggressive and rarest forms of plasma cell dyscrasias. It can arise from a leukemic transformation of multiple myeloma or can be primary and the prognosis is very poor. We present the case of a 68 year old male who presented to our hospital with features of acute renal failure. He was diagnosed as a case of Plasma cell leukemia and was treated with chemotherapy. However his condition deteriorated and he died 2 weeks after diagnosis. The recommended treatment of plasma cell leukemia is aggressive chemotherapy followed by stem cell transplantation. However, once diagnosed, the prognosis is very poor.

**Key words:** Plasma cell dyscrasia, plasma cell leukemia, multiple myeloma.

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### INTRODUCTION

Plasma cell leukemia (PCL) is a rare and aggressive plasma cell dyscrasia. PCL can be divided into primary and secondary types, the latter typically occurring at a late and advanced stage of Multiple myeloma (MM). The prognosis of primary PCL has generally been dismal with reported median overall survival (OS) below 1 year [1]. With the use of novel treatments and autologous stem cell transplantation (ASCT), this has improved somewhat, although the prognosis remains poor.

The diagnostic definition of PCL has traditionally been based on Kyle's criteria from 1974 [2]. In this seminal paper, PCL was defined by at least 20% circulating plasma cells and a total plasma cell count in peripheral blood of at least  $2 \times 10^9/l$ , thereby identifying a leukemic subtype of MM with a particularly poor prognosis.

### CASE PRESENTATION

A 68 year old man presented to the General medicine department with a one week history of breathlessness, facial puffiness and decreased urine output. On examination he had bilateral pitting pedal edema and raised JVP. There was no organomegaly or lymphadenopathy.

Initial laboratory investigations revealed Hemoglobin of 11.6 g%, high WBC count-29800 cells/cu.mm, raised ESR -86 mm/hr and Platelet count -

2.34 lakhs /cu.mm. Renal function was impaired with a high serum Urea-114mg % ( 20-40) and highserum Creatinine- 7.9mg % ( 0.7-1.4).Serum LDH was high - 318 IU/L (150-250).

Other investigations : serum Calcium-12(9-11), Uric acid-14.8 mg% ( 3-7), Total protein - 5.8g%( 6-8), Albumin-2.7g % (3.5-5), Globulin-3.1g%(2-3.5), A/G ratio-0.8(1.4-1.7).

Urine BenceJonce protein-negative

Serum protein Electrophoresis showed absence of M Band. Immunofixation electrophoresis could not be done due to lack of resources.Peripheral smear of the patient showed 22% plasma cells. Bone marrow aspiration was a dry tap. Bone marrow biopsy showed sheets of plasma cells with a few Plasmablasts .Immunohistochemistry showed CD138 positivity and Kappa light chain restriction.

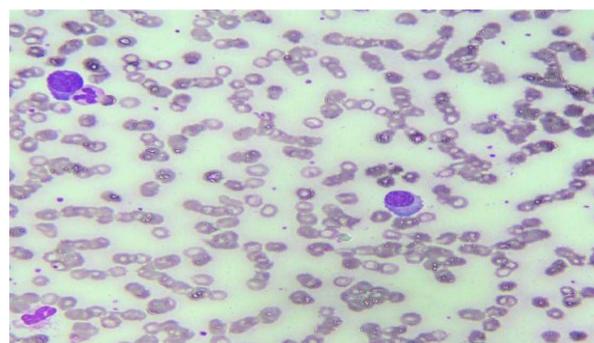
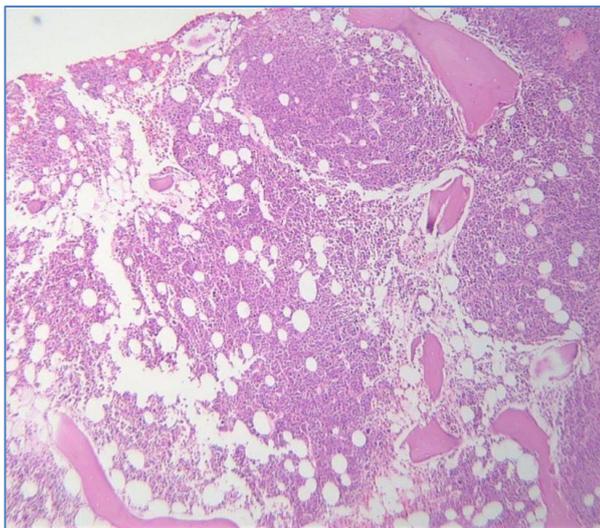
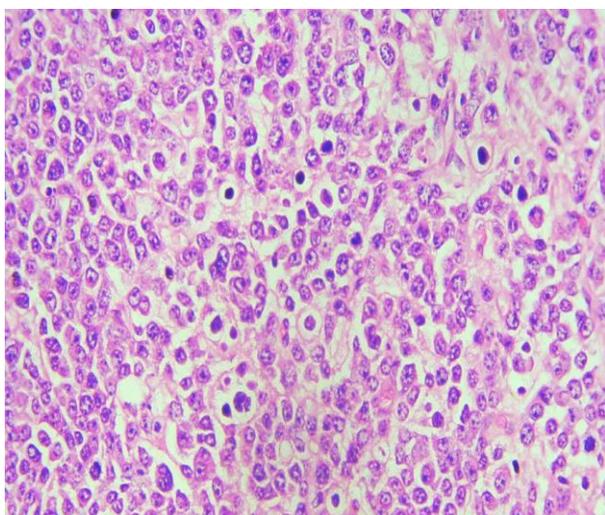


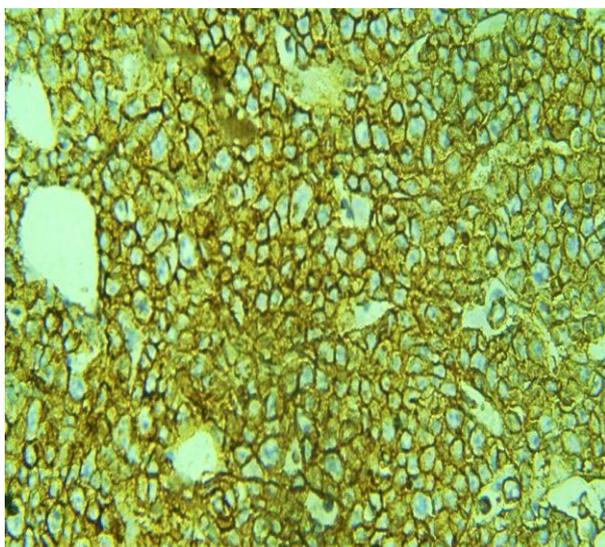
Fig-1: Peripheral smear showing plasma cells



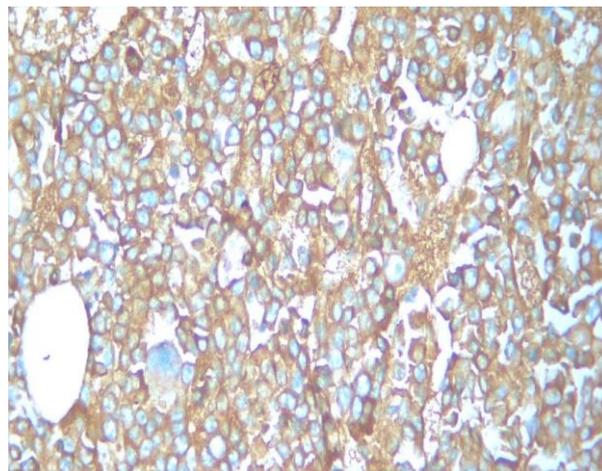
**Fig-2: Bone marrow trephine biopsy – hypercellular marrow**



**Fig-3: High power view of the marrow biopsy showing diffuse infiltration of plasma cells and occasional plasmablasts. Normal marrow elements were not there.**



**Fig-4: The neoplastic cells are CD 138 positive**



**Fig-5: Neoplastic cells show Kappa restriction**

## DISCUSSION

PCL is characterized by the presence of >20% of plasma cells in peripheral blood or absolute plasma cell count  $>2 \times 10^9/L$  [3]. A recent publication suggests that if the diagnostic criteria were reduced to 5% plasma cells and/or an absolute count of  $>0.5 \times 10^9/L$  while incorporating ancillary techniques, patients could be diagnosed earlier and have a better prognosis [4].

Primary PCL has a more aggressive clinical presentation than MM including a higher tumor burden. Patients may present with symptoms due to profound anemia, hypercalcemia, or bleeding diathesis owing to thrombocytopenia. On physical examination, patients may exhibit a higher prevalence of organomegaly with involvement of the liver, spleen, lymph nodes, pulmonary findings associated with pleural effusions, neurological deficits due to central nervous system involvement, pallor, petechiae and palpable extramedullary soft tissue plasmacytomas [5]. Our case had anemia, leukocytosis, elevated serum LDH, hypercalcemia and acute renal failure with elevated creatinine and blood urea. Similar features were also seen in a case series by Rajeshwari et al. [4].

PCL is more frequent in light chain only, IgE and IgD myeloma and is less frequently seen in IgG or IgA myeloma [3]. Our case had absence of M band in Serum electrophoresis and hence we suspected a non secretory myeloma or a light chain only disease. Immunohistochemistry showed a kappa restriction. With the characteristic peripheral smear, bone marrow and IHC findings a diagnosis of primary plasma cell leukemia was considered.

## CONCLUSION

PCL belongs to a unique subset of plasma cell dyscrasias and has both a different biologic background as well as a distinct clinical and laboratory profile. The prognosis of primary PCL is very poor with a median overall survival of only 7 months with standard chemotherapy, and therefore, requires innovative

treatment approaches incorporating various modalities to improve the outcome.

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