Case Report

Primary Plasma Cell Leukemia

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ABSTRACT

Plasma cell leukaemia (PCL) is a rare disease entity where the number of clonal plasma cells in the peripheral blood exceeds 2X109/l or 20% of the total leukocyte count. This plasma cell leukaemia can be primary which is arising DE novo or secondary as in the course of Multiple Myeloma. We report a 50 year male patient who presented with low grade fever, generalised weakness, generalised lymphadenopathy and, subsequently turned out to be primary Plasma cell leukemia after investigations.

Key words : Primary Plasma cell Leukaemia, Multiple myeloma

Introduction :

Plasma cell leukaemia (PCL) is a rare disease entity where the number of clonal plasma cells in the peripheral blood exceeds $2X10^9$/l or 20% of the total leukocyte count[1]. It is classified as either Primary Plasma cell leukaemia occurring at diagnosis or as secondary plasma cell leukaemia in patients with relapsed/refractory myeloma[2]. The clinical course is aggressive with short remissions and survival duration. It is more common in African Americans than in Caucasians[2]. It's common clinical presentation is Hepatosplenomegaly, lymphadenopathy, and fewer lytic bone lesions[3]. Plasmacell leukaemias havepoor response to therapy. We report a case of Plasma cell leukaemia.

Case report

A 50 year old male presented to the outpatient department with Generalized weakness, low grade fever of 1 month, duration & Purpuric rash over both lower limbs 7 days prior to hospitalization. His physical examination revealed stable vitals. He was pale & had purpuric rash over both lower limbs extending up to knees. He had bilateral Cervical, Axillary and Inguinal lymphadenopathy, size varying from 1cm to 3 cm, firm nonmatted & not fixed to underline structure. There were no sinuses. Tere was no sternaltenderness. His systemic exam did not show any abnormalities.

On investigation complete blood count showed increased total Leukocytic count & low platelets. DLC showed predominantly plasma cells, most of which were immature (plasma blasts) around (28%). Cells of neutrophil series were seen with shift to left. ***(Table 1)*** Peripheral smear showed hypochromia & rouleaux formation ***(Fig.1).*** X-ray chest / skull / spine were normal. USG Abdomen / Pelvis showed multiple periportal lymph node (largest 2.3 X 1cm), mesenteric lymphadenopathy & moderate splenomegaly. Bone marrow aspiration revealed large numbers of diffuse population of plasma cells at various stages of maturation. Cells of Erythroid series were few. Myeloid series cells showed normal maturation. Megakaryocytes were normal. ***(Fig. 2)*** Fine needle aspiration cytology from lymph node showed dispersed population of predominantly small to medium sized plasma cytoid cells with scanty moderate amount of cytoplasm with round-oval hyper chromatic nuclei with indentations, clumped chromatic. Histopathological study of Cervical lymph node biopsy showed extensive infiltration of mature and immature plasma cells and Lymphocytes, few binucleated and multinucleated giants cells. ***(Fig. 3)*** Patient's serum protein electrophoresis revealed M band in gamma globulin region (2.4 g/dl) ***(Table 2)*** Flow cytometric of BMA analysis was suggestive of Primary plasma

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cell leukemia. ***(Table 3)***

Though the clinical presentation in this case was unusual a diagnosis of primary Plasma cell leukaemia was entertained on basis of all investigations and confirmed by flow cytometry Patient was treated under haematologist at Mumbai on his request so could not be followed further.

Table 1 **- Results of complete blood count**

Investigation	**Result**
Haemoglobin	9.1g/dl
Total leucocyte count	16100/cumm
Differential count	
Neutrophil	64%
Lymphocyte	33%
Monocyte	2%
Eosinophil	1%
Atypical cell (plasma)	28%
Platelet count	61000/cumm

Table 2 **- Results of Serum electrophoresis**

Investigation	**Result**
Total serum protein	9.89g/dl
Albumin	2.4g/dl
Alfa 1	0.3g/dl
Alfa 2	0.5g/dl
Beta 1	0.3g/dl
Beta 2	0.9g/dl
Gamma globulin	5.6g/dl
M band	Detected in gamma globulin region 2.4g/dl
Albumin/Globulin	0.32

Table 3 : **Flow cytometry of bone marrow aspiration**

Investigation	**Result**	**Positive**	**Negative**
Flow cytometry of bone marrow aspiration	29%polyclonal plasma cell	CD38,CD138, CD19, CD45	CD56,CD20,CD117
	kappa & lambda ratio 1:1		

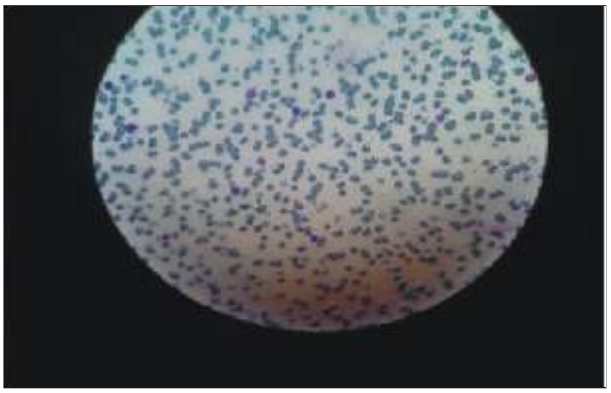

Figure 1 : **Leishman`s stained peripheral smear : high power view showing more than 20% plasma cell (magnification 40x)**

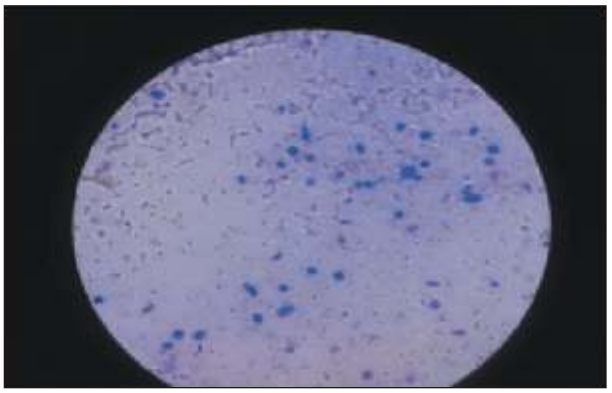

Figure 2 : **May Grunwald-Geimsa stained bone marrow aspiration smear : high power view showing plasma cell & myeloid series cells (magnification 40x)**

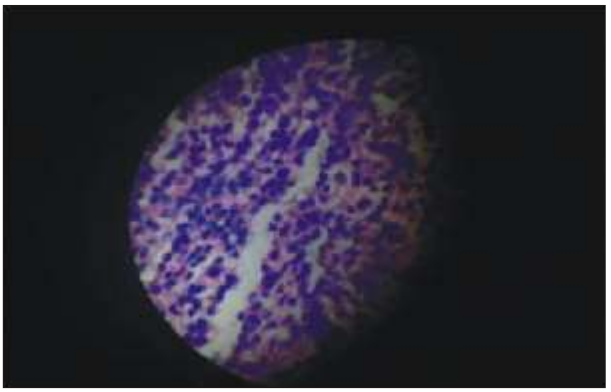

Figure 3 : **Haematoxylin & Eosin stained smear from cervical lymph node biopsy showing mature & immature cells**

Discussion :

Primary plasma cell leukaemia is important to recognize because of its rarity of occurrence and unusual presentation. The incidence of Secondary plasma cell leukaemia occurs as a progression of the disease in 1% to 4% of all cases of myeloma. The incidence of primary plasma cell leukaemia is very rare and reported to occur in less than one in a million[1]. Primary plasma cell leukaemia has been reported to be associated with prior exposure to chemotherapy and/or radiotherapy; however, this association is difficult to confirm due to low incidence of the disease[4]. This case is presented because of its rarity & presentation at a younger age. Primary plasma cell leukaemia has more extensive bone marrow involvement ,a higher prevalence of organomegaly with involvement of the liver, spleen & lymph nodes. Renal failure is more common. However bone involvement (lytic lesions) is rare. Response to therapy is poor.

Immunophenotypically, these clonal plasma cells express either κ or γ light chains, but not both and demonstrate increased expression of CD138, CD38 and CD20 (especially primary plasma cell leukaemia) & lack CD56, CD9, CD117 and human HLA DR expression. Lack of CD56 expression could explain the propensity of these clonal plasma cells to leukemic transformation and also account for the lower osteolyticpotential and higher rates of extra medullary involvement seen in patients with plasma cell leukaemia[5].

In plasma cell leukaemia Intensive chemotherapy regimens and bortezomib-based regimens (or Thalidomide / Linolidamide) are recommended followed by high-dose therapy with autologous stem cell transplantation if feasible. Allogeneic transplantation can be considered in younger patients. Bortezomib based therapy rapidly reduces tumour load and reverses complications, including renal failure and hypercalcemia.[6]

Overall prognosis of Plasma cell leukaemia remains poor. With conventional chemotherapy the median survival is estimated to be 7 months and even with autologous or allogeneic stem cell transplant, survival is usually not longer than 3 years. Secondary plasma cell leukaemia has a particularly poor prognosis, with an estimated median overall survival of approximately 2 months.[5]

Conflicts of interest : None reported by Authors

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