Primary plasma cell leukemia: A rare case report

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ABSTRACT

Plasma cell leukemia (PCL) is a rare form of plasma cell dyscrasia. Between 2% and 4% of malignant PCD cases are PCL. The presentation may be primary, de novo, or secondary, evolving from an existing case of myeloma as part of the terminal phase of the disease. The median age of patients is 50-60 years with an equal incidence in males and females. Here, we report a case of primary PCL, presenting at the age of 62 years.

Keywords: *Leukemia, Plasma cell, Plasma cell dyscrasia.*

The first case of plasma cell leukemia (PCL) was recognized by Gluzinski and Reichentein [1]. The presentation may be primary, de novo, or secondary, evolving from an existing case of myeloma as part of the terminal phase of the disease. About 60% to 70% of cases are primary [2]. The incidence of primary PCL is very rare and reported to occur in less than one in a million [3]. PCL patients usually have accompanying anemia, hypercalcemia, renal insufficiency and organomegaly. Two types of PCL are seen: secretory and non-secretory. No M-protein is detected in the non-secretory type of PCL. PCL is more frequent in the light chain only, IgE and IgD myeloma and is less frequently seen in IgG or IgA myeloma.

By definition, there are more than 20% plasma cells in the peripheral blood and an absolute plasma cell count of more than $2x10^9$/L [4,5]. PCL has a relatively poor prognosis, due to its very aggressive nature involving extramedullary organs, lytic bone lesions and bone marrow failure. Treatment includes immunomodulators, proteasome inhibitors, and autologous stem cell transplantation. Outcomes are not promising, however, even after treatment; median survival after chemotherapy and transplant is not more than three years [6]. As prognosis is very poor, it is very important to recognize this entity sufficiently early so that one can offer combination chemotherapy at the earliest which can prolong survival.

CASE REPORT

A 62-years-old female presented to the emergency with complaints of weakness, easy fatigability, back pain, and high-grade fever since two months. She did not have any significant past and family history. On examination, the vitals were stable. The patient was investigated.

A complete hemogram showed haemoglobin of 7.7 g/dl, total white blood cell count of 8000/mm³ and a platelet count of 50,000/mm³. Peripheral smear showed 26% of immature and mature plasma cells (absolute count=2,080/mm³) and numerous reactive lymphocytes. Rouleaux formation was also seen (Fig. 1).Renal function tests were deranged with blood urea levels of 315mg/dl and serum creatinine levels of 7.5 mg/dl, and serum uric acid levels of 10mg/dl. Ultrasound revealed hepatosplenomegaly. Urine was negative for Bence Jones Proteins. Serum electrophoresis was done. High-resolution serum protein electrophoresis revealed the presence of a densely staining monoclonal gammopathy ('M' spike) in the gamma globulin region. A bone marrow aspiration was then done. Bone marrow smears revealed diffuse involvement of bone marrow by mainly mature plasma cells (86%) and few immature plasma cells (9%) (Fig. 2).

Based on the findings of peripheral smear andbone marrow,the patient was diagnosed to have pPCL. He was given supportive care and bortezomib based chemotherapy was initiated. However, the patient was lost to follow-up.
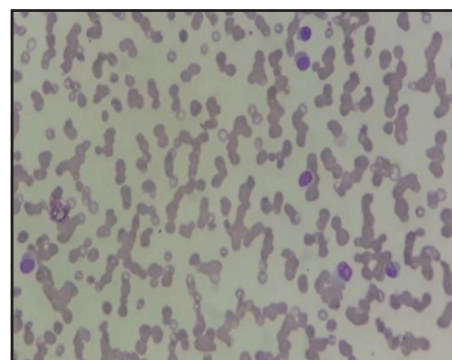

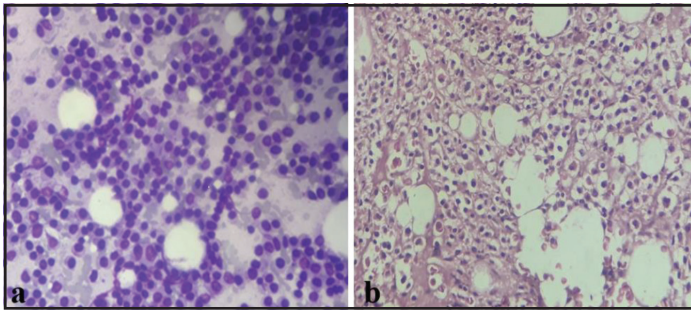**Figure1: Peripheral smear showing Rouleaux formation and plasma cells (Leishman stain, 400X).**

Figure 2: (a) Bone marrow aspirate showing plasma cells (Leishman stain,400X); (b) bone marrow biopsy showing sheets of plasma cells (H& E stain, 400X).

DISCUSSION

Plasma cell leukemia is a rare and aggressive variant of plasma cell dyscrasia, accounting for only 2-4% of all plasma cell dyscrasias [7]. By definition, there should be >20% plasma cells in the peripheral blood or an absolute plasma cell count $>2 \times 10^9$/L.The median age of patients is 50-60 years with an equal incidence in males and females. Phenotypically, they originate from the proliferation of plasma cells expressing CD38 [8] .

Clinical presentation of Plasma cell leukemia is more aggressive than that of multiple myeloma with a higher presenting tumour burden and higher frequencies of extramedullary involvement, anemia, thrombocytopenia, hypercalcemia, renal impairment, increased levels of serum lactate dehydrogenase, beta-2 microglobulin and plasma cell proliferative activity [9]. Though the clinical and laboratory features of primary and secondary plasma cell leukemia are similar, the response to therapy and overall survival in primary plasma cell leukemia goes from poor to worse.

Response to treatment of PCL is poor. Median survival is less than 1 year.Since the prognosis is so poor, intensification of high dose chemotherapy followed by allogeneic/autologous stem cell rescue should be tried [10]. The most important prognostic factor in pPCL remains response to treatment as patients presenting with the disease that is resistant to initial therapy have the poorest outcome [11].

The current treatment plan follows the steps involved in the management of multiple myeloma. Induction therapy includes various bortezomib-based regimens such as VDT-PACE (bortezomib, dexamethasone, thalidomide, cisplatin, adriamycin, cyclophosphamide, and etoposide), VDT (bortezomib, thalidomide, and dexamethasone), VAD (bortezomib, doxorubicin, and dexamethasone), VRD (bortezomib, lenalidomide, and dexamethasone) or VMP (bortezomib, melphalan, and prednisone).

Although the best induction regimen for PCL is not known. Recently, lenalidomide and bortezomib-based regimens have demonstrated activity and are more widely used. In a multicenter retrospective study involving 42 patients with pPCL, bortezomib-based therapies were associated with 69% response rates,

1–3-month median survival time [12]. Although there is great variability in treatment plans, typically individuals <65 years in good performance status are treated with aggressive induction therapies such as VDT-PACE.

CONCLUSION

Plasma cell leukemia is a rare and aggressive form of leukemia with poor prognosis. As survival rates are low, such information is necessary to ensure PCL cases can be diagnosed early and appropriate treatment regimens initiated.

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