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Case Report

IgA Plasma Cell Leukemia

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ABSTRACT

Plasma cell leukemia (PCL) is a rare entity. There are two presentations of PCL, primary or secondary. The primary or *de novo* form of PCL presents with an acute and rapidly progressive leukemic phase. This form occurs when the patient has no pre-existing multiple myeloma (MM). The secondary form is the most advanced form of MM. The PCL is a rare disorder representing 1–2% of the diagnosed cases of MM. Median age at presentation is usually above 50 years. The monoclonal protein in patients with PCL may be IgG (50%), IgA (15%), or in rare cases IgD or IgE (6%). We report a case of IgA primary PCL that is very rare. Patient was started on combination therapy with vincristine, adriamycin, and dexamethasone. There was poor response and patient died three months after diagnosis.

Keywords: Primary plasma cell leukemia, multiple myeloma, plasma cell dyscrasia

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INTRODUCTION

Plasma cell leukemia (PCL) is a rare form of plasma cell dyscrasia characterized by proliferation of plasma cells that account for more than 20% of the differential WBC count. The concentration of plasma cells exceeds 2×10^9/l.[1] The primary form presents *de novo* in patients with no previous history of multiple myeloma (MM) and usually features a rapid clinical progression and short survival. The secondary type evolves as a terminal event in 1–2% of MM. The clinical presentation of PCL differs from MM and resembles that of acute leukemia. The disease manifests itself with weight loss, fatigue, anemia, and bleeding. The median survival is as low as six months. It is very important to recognize this entity sufficiently early so that one can offer combination chemotherapy at the earliest followed by stem cell transplant which can prolong survival.[2] Here, we report a case of primary PCL who was treated with combination of vincristine, adriamycin, and dexamethasone (VAD).

CASE REPORT

A 75-year-old male presented with complaints of loss of weight, anorexia, and generalized body ache of four weeks duration. On examination, he was pale with upper cervical nontender lymphadenopathy, tachycardia, and no organomegaly. Hemogram revealed hemoglobin of 7.6 g/dl, total white cell count of 39.6×10^9/l, and platelet count was 171×10^9/l. Peripheral smear showed hypochromia and anisopoikilocytosis with 46% plasma cells and plasma blasts with roeuleux formation [Figure 1]. Other abnormal laboratory studies were high sedimentation rate of 120 mm/h LDH-1130 IU/l, serum creatinine 1.6 mg/dl, and serum calcium 12 mg/dl. Coagulation parameters showed prothrombin time (PT) 16 s, activated partial thromboplastin time (aPTT) 42 s, serum fibrinogen 2.2 g/dl, and FDP 30. Bone marrow aspirate showed 65% plasma cells and plasma blasts [Figure 2]. Liver and renal function tests showed hypoalbuminemia, hyperuricemia, and raised serum creatinine. Serum protein electrophoresis and immunofixation studies showed IgA 2 950 mg/dl, IgG 628 mg/dl, IgM 16.1 mg/dl, and IgA kappa paraprotein at a concentration of 2.95 g/dl. Skeletal survey showed well-defined lytic lesion in right frontal bone. Immunohistochemistry of bone marrow biopsy showed positivity for CD138 and CD38. Other

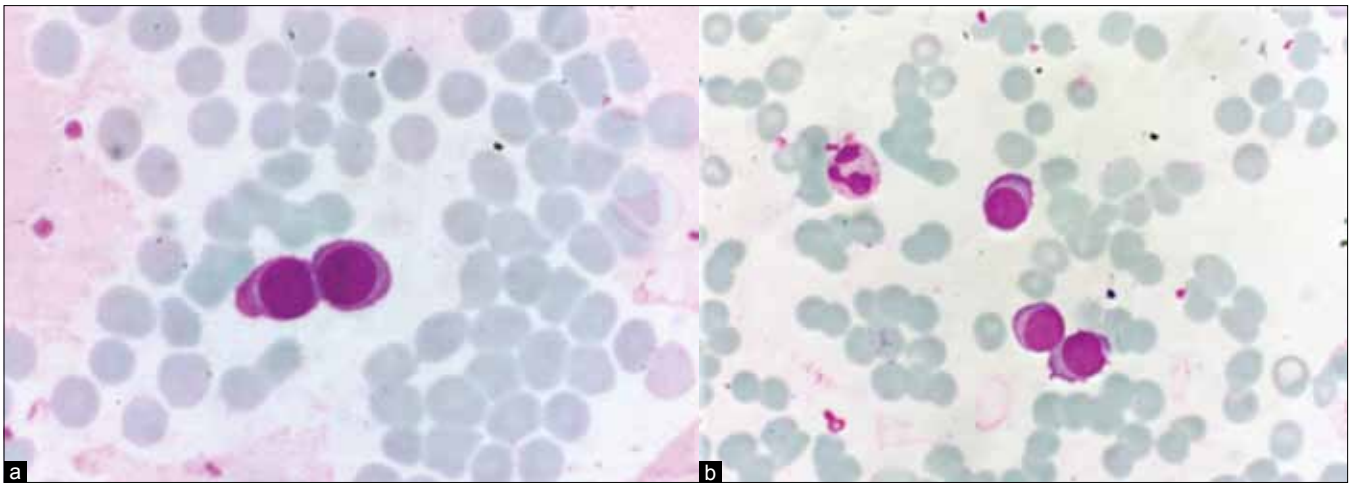

Figure 1 (a-b): Peripheral blood smear showing plasma cells with eccentric nucleus, perinuclear halo with basophilic cytoplasm

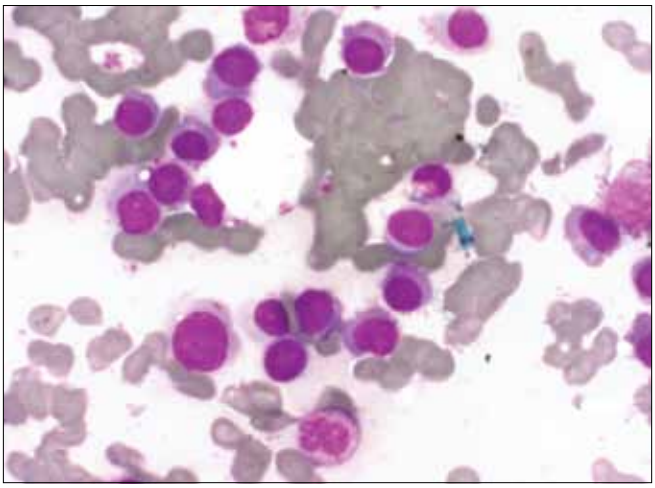

Figure 2: Bone marrow trephine biopsy shows nodular aggregate of neoplastic plasma cells. H&E, x400

markers like CD45, CD56, and CD19 were negative. Kappa light chain was also positive. Based on the above findings our patient was diagnosed to have primary PCL. He was started on supportive care and chemotherapy with VAD regimen. After the first cycle of chemotherapy, plasma cells in peripheral smear decreased to less than 10% and patient showed symptomatic improvement. He received one more cycle of chemotherapy with same regimen, however, the disease progressed and he died after one month due to chest infection and respiratory failure.

DISCUSSION

Although plasma cells are occasionally observed in the peripheral blood of patients with myeloma, the term PCL is only used when the number of these circulating cells is significant (more than 20%).[1] Because of the low frequency of PCL, most clinical data are collected from case reports and few reviews. Its incidence ranges from 1 to 2% of all myelomas.[1,3,4] Phenotypically they originate from proliferation of plasma cells expressing CD38, minority of cells express CD10, HLA DR, and CD20. The presence of multiple hemopoietic surface antigens in malignant plasma cells suggests its origin from pluripotent stem cell. Primary PCL shows higher expression of CD20 as compared to MM. Also, plasma cells from both primary and secondary PCL lack CD56.[5] Among immunoglobulins, IgG is most often increased. Few cases of IgA PCL have been reported.[6] Response to treatment of PCL is poor. Median survival is less than one year. The longest survival reported was 28 months.[7] The failure to achieve 50% clearance of blood plasma cells within 10 days after the initiation of treatment is a predictor of no response.[8] Treatment with a single alkylating agent plus prednisolone is not appropriate for patients with PCL. Combination chemotherapy with VAD, cyclophosphamide, and etoposide or other regimens seems to be a better modality.[8] Our patient showed no improvement with VAD chemotherapy with less than 10% clearance of blood plasma cells within 10 days of starting treatment. Because of the poor prognosis of PCL, intensive chemotherapy and subsequent consolidation with autologous or allogenic stem cell transplantation could be a better approach especially in younger patients who are in good clinical condition.[2]

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