Case Report

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A case report of a young female with plasma cells leukemia

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Abstract:

Plasma cell leukemia (PCL) is a rare and aggressive variant of plasma cell dyscrasias that can occur either *de novo* (primary) or as a leukemic transformation of end-stage multiple myeloma (MM). PCL is known with a poor prognosis and with a different biologic background, clinical, and laboratory features. Primary PCL presents more often with extramedullary involvement, anemia, thrombocytopenia, hypercalcemia, elevated serum 2-microglobulin, and lactate dehydrogenase levels, in addition to impaired renal function in comparison with MM. Here, we report a case of a young female who was admitted with excessive thirst, polyuria, lower backache, fever, and weight loss over 2 months. Peripheral blood smear showed numerous atypical looking plasma cells, and immunophenotyping on bone marrow (BM) aspirate demonstrated the presence of clonal plasma cells. She was admitted to adult hematology ward in Hiwa Hemato-Oncology Hospital, Sulaimani, Kurdistan, Iraq and received four cycles of bortezomib, thalidomide, and dexamethasone chemotherapy, followed by two more cycles of dexamethasone, cyclophosphamide, etoposide, and cisplatin, and later, she proceeded to autologous BM transplantation. More details of the case are presented below.

Keywords:

Autologous transplantation, leukocytosis, plasma cell leukemia

Introduction

Primary plasma cell leukemia (pPCL) is an aggressive plasma cell proliferation. It is defined by the presence of >2 × 10^9/L circulating plasma cells or plasmacytosis of >20% of the differential white count, in the absence of preexisting multiple myeloma (MM).[1,2] The first report of plasma cell leukemia (PCL) was in 1906 by Gluzinski and Reichenstein in a patient who presented with bone pain, a palpable rib mass, rib fractures, anemia, and splenomegaly.[3,4] Although PCL and MM are closely related disease entities, the prognosis of PCL patients treated with standard chemotherapy has consistently shown to be inferior.[5,6] In pPCL, clonal plasma cells accumulate in the bone marrow (BM) and also have an enhanced capacity to recirculate in blood, with subsequent formation of extramedullary disease.[4] The dissemination of tumor cells out of the BM is not only related to variation in the expression of adhesion molecules and chemokine receptors but also to the presence of other molecular aberrations, which contribute to BM microenvironment-independent tumor growth, suppression of apoptosis, and the escape from immune surveillance.[7,8] These genetic abnormalities are already present at diagnosis of pPCL.[9,10] The outcome of pPCL has much improved after the introduction of autologous stem cell transplantation and combination approaches with novel agents, including bortezomib and immunomodulatory drugs, such as lenalidomide.[4,10]

Case Report

A 36-year-old female who presented to our hospital with 2-month history of excessive thirst, polyuria, generalized bone aches, particularly in the lower back, fever, and weight loss. Physical examination showed

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scattered ecchymosis with bilateral cervical and submandibular lymphadenopathy.

The laboratory evaluation on admission showed hemoglobin (Hb) of 8.1 g/dl, white blood cell count of 29×10^9/L, platelets count of 11×10^9/L, and erythrocyte sedimentation rate of 70 mmHg/h. The blood smear demonstrated leukocytosis with numerous atypical plasma cells that amount to 60% of the differential count [Figure 1]. Immunophenotyping performed on bone marrow aspiration (BMA) sample and revealed the presence of clonal plasma cells (CD38+, CD138+, CD27+, Kappa+, CD45−, CD19−, CD20−, CD117−, and CD56−) comprising about 95% of all BM nucleated cells [Figure 2].

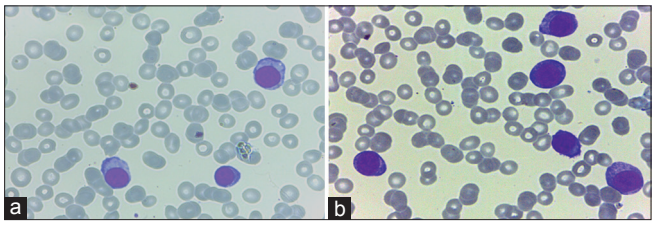

Figure 1: (a and b) Circulating atypical plasma cells in peripheral blood

Additional investigation returned with a serum calcium level reaching 12 mg/dL, lactate dehydrogenase (LDH) of 668 U/L, B2-microglobulin of 1.8 mg/dL, albumin of 5 g/dL, and no M-band detected on serum protein electrophoresis. While an excessive light chain protein excretion was reported in urine protein electrophoresis, urine protein was 16.1 g/dL, and urine immunofixation confirmed the presence of free Kappa light chain with negative urine for Bence Jones protein. The other investigations were normal including renal function test. The skeletal survey showed no lytic lesions.

The patient was admitted to adult hematology ward in Hiwa Hemato-Oncology Hospital, Sulaimani, Kurdistan, Iraq and received supportive therapy with proper hydration, allopurinol, antimicrobial prophylaxis, zoledronic acid, blood, and platelets transfusion. Treatment with bortezomib, thalidomide, and dexamethasone (VTD) chemotherapy protocol was initiated; the general condition of the patient was improving with rising Hb and platelets count with gradual resolution of leukocytosis. However, circulating plasma cells were still detected on the blood smear. After four cycles of VTD, the investigations showed normal blood counts, peripheral blood smear, and normal

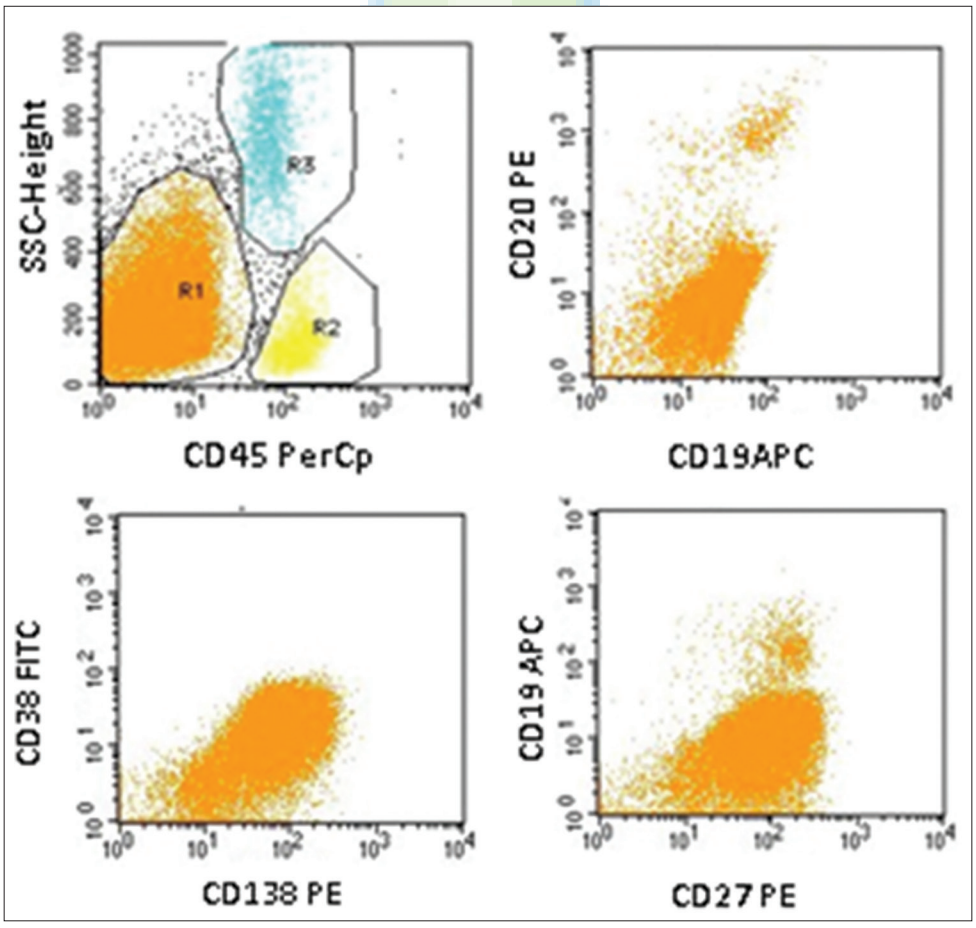

Figure 2: The atypical plasma cells (R1) that are CD45−, CD19−, CD20−, CD 38 weak+, CD138+, and CD27+

serum LDH with an excellent performance status of the patient, unlike the BMA that showed 51% of plasma cells. After that, two more cycles of dexamethasone, cyclophosphamide, etoposide, and cisplatin were begun. The BMA at that stage demonstrated about 6% of the plasma cell. She proceeded to autologous BM transplantation, after conditioning regimen with 200 mg/m^2 of melphalan. The early posttransplantation course was uncomplicated except for an episode of bloody diarrhea and pancytopenia that developed at 3 and 5 weeks, respectively. However, the patient did not have a BM relapse. A polymerase chain reaction confirmed high cytomegalovirus viral load, and thus, ganciclovir was initiated. Subsequently, the patient achieved complete medical remission. Should she relapse after autologous BM transplant, the care plan will be to arrange for human leucocyte antigen-identical allergenic BM transplantation when a compatible donor is available.

Discussion

PCL is a highly aggressive disease, with no consensus on a standard chemotherapy regimen because of the condition rarity. PCL comprises less than one patient/1000,000 populations.[10] Hiwa hospital provides cancer service for more than 2000,000 populations. Despite that, we reported only three cases of PCL admitted to our hospital over the past 10 years; the first two cases died early within the first few months of presentation due to the lack of novel PCL therapy agents at that time. The condition of this young female has illustrated the aggressive clinical course of pPCL. She presented with typical features of the disease: excessive thirst, polyuria, generalized bone aches, fever, and weight loss over 2-month duration. Moreover, she had poor prognostic signs of a grave outcome (hypercalcemia and high LDH). In this case, we also demonstrated the significant role of flow cytometry in the diagnosis of clonal plasma cell dyscrasias, and we overviewed the currently available therapeutic options at our hospital and the plan for such patients in case of relapse.

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Conflicts of interest
There are no conflicts of interest.

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