Case Report

Abrupt Development of Plasma Cell Leukemia in a Patient with Chronic Anemia under Follow-up

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Abstract

Plasma cell leukemia (PCL) is a very rare and progressive hematologic malignancy with unpleasant prognosis, which present with monoclonal proliferation of plasma cells in peripheral blood. Here we report a 52-year-old female case of PCL which diagnosed by morphology and immunohistochemistery (IHC) study. IHC revealed CD20+/CD38+/CD138+/CD56-/kappa-/lambda+. We diagnose PCL on peripheral blood. IHC can be helpful for prognostic determination.

Keywords: Abrupt, chronic anemia, leukemia, plasma cell

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Introduction

Plasma cell leukemia (PCL) is a very rare and aggressive malignant neoplasm arises from bone marrow that present with many plasma cells in the peripheral blood. PCL can be primary or may be arise from multiple myeloma.[1] Clinical manifestation is resembled to multiple myeloma, such as anemia, bone pain, renal involvement and hypercalcemia. Peripheral smear may be the first diagnostic step with at least 20% plasma cell of all white blood cells.[2] Malignant plasma cells may have very different configuration from normal plasma cells to cells that are often barely recognizable as being plasma cells.[3] Other laboratory finding in PCL is protein electrophoresis and lytic lesion in skull X-ray. Immunophenotyping show positive reaction against CD38, CD138 and CD56, but negative for CD2, CD3 and CD16. The survival rate is very short and less than a year. In one study deletion of 13q was very common by the FISH test.[1] Some other malignant disease such as angioimmunoblastic T-cell lymphoma revealed somehow peripheral plasmacytosis that mimicking plasma cell leukemia.[4]

Case Report

A 52-year-old female presented with generalized bone pain and pallor for several months. General physical examination showed chronically ill patient with bone pain without organomegaly. She was under follow up for seven years as chronic anemia.

Complete blood count revealed 110000 white blood cell count and anemia (with hemoglobin 8 mg/dL), as well as platelet count less than 100000. ESR was 96, although previous hematologic results only showed anemia without any evidence of malignancy.

Peripheral blood smear showed more than 90% plasma cells (Figure 1). Blood urea nitrogen was 41 mg/dL and creatinin 1.4 mg/dL and also liver enzymes, especially LDH was more than 1400 u/L. Patient referred to an oncologist with presumptive diagnosis of PCL; therefore bone marrow biopsy was done and showed hypercellular marrow packed with plasma cells (Figures 2 and 3). Protein electrophoresis revealed M component. Immunohistochemistry study revealed positive reactions for CD138, CD38, Lamda light chain, and CD20 antibody but negative for CD56 and Kappa light chain antibody (Figures 4 – 6).

Skull X-ray showed no evidence of lytic lesion. The ultrasound study didn't show organomegaly. After a good response to chemotherapy result of laboratory tests were excellent that complete blood count and bone marrow examination were cleared from malignant plasma cells. She was a candidate for a bone marrow transplant, however, she expired due to renal failure

Discussion

We report a rare case of hematologic neoplasm. The patient abruptly developed PCL and presented with chronic anemia for several years. Tiedmann, et al. noticed that the median age of PCL cases is 55, about a decade younger than the median age of multiple myeloma patients.[5] IHC from the bone marrow biopsy support our diagnosis that expressed CD38/ CD20/ CD138/ lambda chain and negative for kappa chain and CD56.

IHC is very helpful in diagnosis of PCL specially for atypical forms.[1,2,6] Sahara, et al. present cases of multiple myeloma some of which are CD56 negative that have extramedullary involvement and poor prognosis.[7] PCL patients usually developed anemia and severe thrombocytopenia.[5,8,9]

CD56 negative cases of PCL are more susceptible to renal insufficiency.[7] CD56 negative cases are reported in patients with PCL rather than multiple myeloma.[10] CD56 is an adhesion molecule that loss of expression of this molecule explains CNS involvement in aggressive forms of PCL.[11]

Our case did not express CD56 on plasma cells in IHC study that show aggressiveness and susceptibility to extra medullary involvement.

Some researches show that the treatment of PCL is not effective

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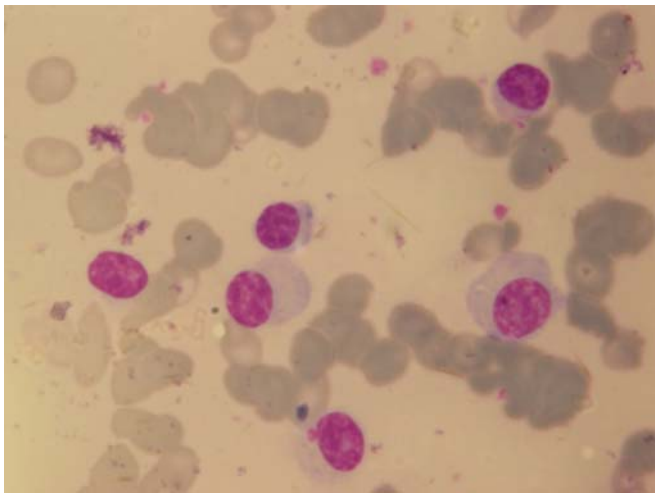

Figure 1. Peripheral smear showing many plasma cells in patient with plasma cell leukemia, Giemsa stain, 1000x.

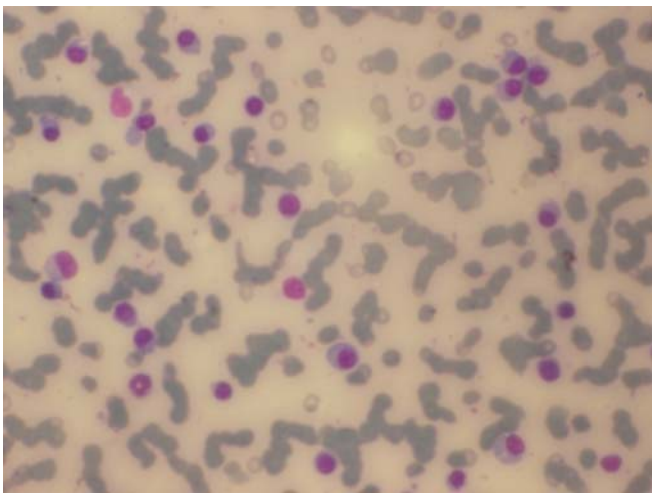

Figure 2. Bone marrow aspiration showing diffuse involvement of bone marrow by typical plasma cells, Giemsa staining, 400x.

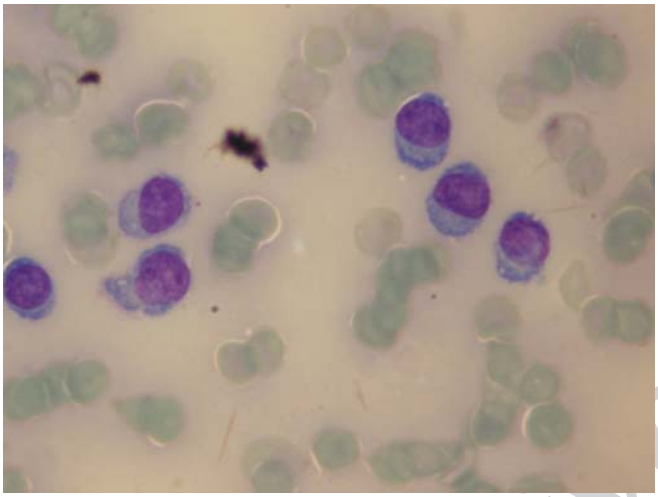

Figure 3. Bone marrow aspiration showing diffuse involvement of bone marrow by typical plasma cells, Giemsa staining, 1000x.

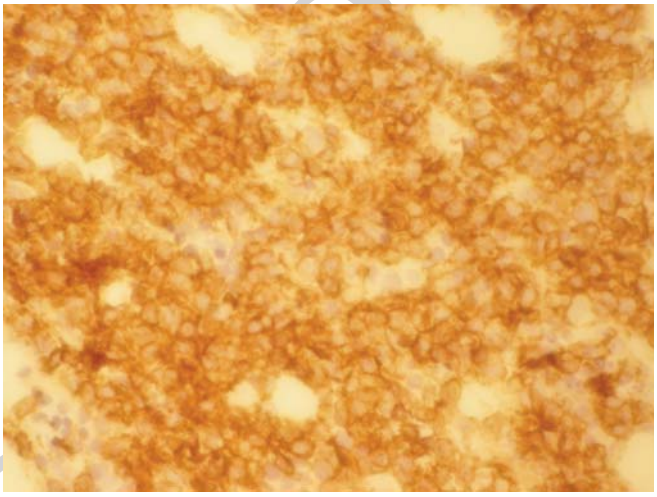

Figure 4. Plasma cell leukemia. Immunohistochemistry on paraffin embedded tissue of bone marrow revealed positive reaction for anti CD 138.

Figure 5. Plasma cell leukemia. Immunohistochemistry on paraffin embedded tissue of bone marrow revealed positive reaction for Lambda chain antibody.

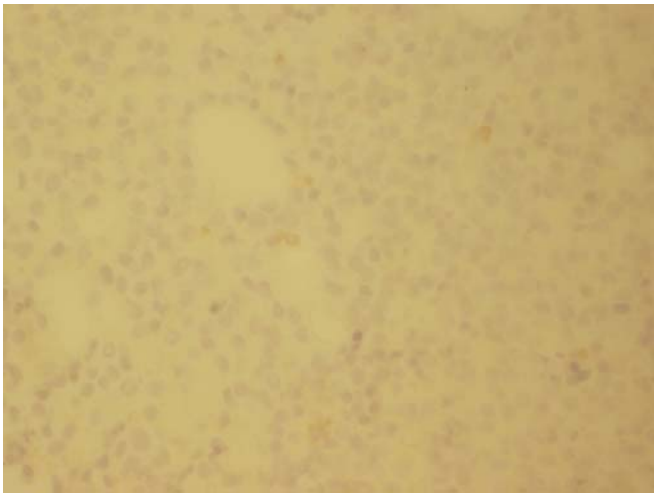

Figure 6. Plasma cell leukemia. Immunohistochemistry on paraffin embedded tissue of bone marrow revealed negative reaction for anti CD 56.

that the survival of these patients is less than 1 month in 28%.[5,9,12] Even though our patient successfully tolerate treatment and after a short period her bone marrow and peripheral smear were cleared from plasma cells.

In summary, according to our case and review of the literature we think that CD56 negative plasma cells are more aggressive and mostly support a diagnosis of plasma cell leukemia.

Conflict of interest: *None declared*

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