

Leukemia with Plasma Cells — Case Report

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ABSTRACT

Plasma cell leukemia (PCL) is one of the most aggressive monoclonal gammopathies, being characterized by the presence of more than 20% of plasma cells in the peripheral blood and an absolute number of these cells of more than 2×10^9 , with different morphology, from young elements to mature cells. The incidence of PCL varies between 2–4% among multiple myeloma (MM) patients. In comparison with MM, PCL appears more often in younger patients. The following article describes the case of a 49-year-old female patient diagnosed with PCL which needed urgent control of the clinical manifestations due to its irreversible complications. Urgent autologous stem cell transplantation is recommended in this group of patients.

Keywords: plasma cell leukemia, multiple myeloma, autologous stem cell transplantation

INTRODUCTION

Plasma cell leukemia (PCL) is one of the most aggressive monoclonal gammopathies, being characterized by the presence of more than 20% of plasma cells in the peripheral blood and an absolute number of these cells of more than 2×10^9 , with different morphology, from young elements to mature cells.¹ The disease can be accompanied by either the presence or the lack of immunoglobulin secretion, with or without bone lesions.

The incidence of PCL varies between 2–4% among multiple myeloma (MM) patients. In comparison with MM, PCL appears more often in younger patients.² The median age of patients diagnosed with PCL is 55 years, ten years lower than the median age of MM patients at diagnosis.

PCL can appear as a primary, “de novo” disease in patients who have not been in hematological evidence with multiple myeloma before, or as a secondary affection — the leukemic transformation of a formerly known relapsed or refractory MM.³

The clinical presentation of PCL at the beginning can be similar to MM, with the presence of anemic syndrome, which is more frequent at the start of the disease and is usually more severe.⁴ The presence of hemorrhagic syndrome, because of thrombocytopenia, is more frequent in PCL patients. Renal dysfunction can be observed in approximately 50% of MM patients, 20% of them de-

veloping renal failure. Up to 50% of the newly diagnosed patients present decreased creatinine clearance, and approximately 9% need dialysis due to the severe deterioration of renal function.⁵

Laboratory findings are similar to those in MM patients, with a couple of exceptions: higher frequency of thrombocytopenia and anemia, and the peripheral smear contains more than 20% of plasmocytes, an essential element for the diagnosis of the disease. Hypercalcemia and renal dysfunction appear frequently; while osteolytic lesions are less expressed and extended as in MM, lactate dehydrogenase and beta-2 microglobulin levels are highly elevated.^{6,7}

PCL needs urgent control of its clinical manifestations in order to prevent early mortality due to its irreversible complications. Urgent autologous stem cell transplantation is recommended in this group of patients.⁸

CASE PRESENTATION

We present the case of a 49-year-old female patient, with urban provenience, known from her personal pathological history with operated right renal lithiasis, who presented at the doctor's office in September 2016 with the following symptoms: altered general status, physical asthenia, fatigue, predominantly inspiratory dyspnea, diffuse bone pain, and oligoanuria. The clinical examination revealed pale skin and mucous membranes, abolished bilateral basal vesicular sound, tachycardia (116 beats/minute), holosystolic murmur caused by tricuspid insufficiency with regurgitation, abdomen increased in volume, liver 3 cm under the right costal margin and impalpable spleen. The patient needed erythrocyte mass transfusion for the correction of the anemia.

The bone marrow aspirate showed: rich cellularity, diffuse infiltration with atypical plasmocytes with myelomatous character, relative frequency of plasmoblasts (plasmocytes with visible nucleus) in proportion of approximately 60%. There were plasmocytes with 2–3 nuclei with cytoplasmic vacuolation, cytoplasmic extensions associated with a lymphoplasmacytic population with reniform nucleus and cytoplasmic extensions (similar to hairy-cell leukemia cells). Slightly reduced cellular lines, E/G ratio: 1/2. Relatively well represented erythroid and granulocytic series with normal aspect and good maturation, as well as rare megakaryocytes with the presence of nucleus were described.

The peripheral smear revealed the presence of plasmocytes in a proportion of 25%, some of them presenting cytoplasmic extensions, anisocytosis, and poikilocytosis.

In January 2017, peripheral blood immunophenotyping showed 25% of plasmocytes with the following antigenic profile: CD45–, CD19–, CD38+ dim, CD20–, CD117–, CD56+, CD81+, ic Kappa–, ic Lambda+.

Abdominal ultrasound was performed with the following results: liver with homogeneous echotexture, preserved echogenicity, right lobe 160 mm, inferior cava vein 26 mm, without inspiratory collapse, with visible suprahepatic veins until the periphery; partially contracted gall bladder with edematous walls and transonic content; portal vein 13 mm, splenic vein 15 mm, principle bile duct 4.8 mm, non-expanded intrahepatic bile ducts; echogenic pancreas of normal dimension; right kidney 98 mm, parenchymal index (PI) approximately 10 mm, with some cysts and an 8 mm hyperechogenic formation in the medial calyx (suggestive for renal calculus); left kidney 110 mm, PI 14 mm, without image of calculus, without hydronephrosis; homogeneous spleen of 104/40 mm; evacuated bladder, apparently normal uterus of normal dimensions; a layer of liquid accumulation in the costodiaphragmatic sinuses; a layer of liquid accumulation around the liver, gallbladder, and kidney in small/medium quantity. Conclusions: minor portal hypertension, right renal calculus, medium quantity ascites, bilateral pleurisy.

Native computed tomography: multiple secondary osteolytic determinations of the skeleton, on costal levels several osteolytic zones extended into the underlying and overlying soft tissues.

Based on the examinations presented above, we established the diagnosis of acute plasma cell leukemia as a transformation of multiple myeloma, and chemotherapy treatment has been initiated. Until present, the patient benefited of five chemotherapy courses with vincristine, epirubicin, cyclophosphamide, melphalan, and dexamethasone, in association with bortezomib and bisphosphonate remineralizing treatment, followed by establishing an appointment for stem cell mobilization and harvesting.

The patient needed to continue hemodialysis courses for three weeks, followed by the normalization of creatinine, urea, and glomerular filtration rate values.

After the fifth chemotherapy treatment, the patient presented an acute pyelonephritis with multidrug-resistant *Staphylococcus aureus*, with highly increased C-reactive protein (497 mg/L) and procalcitonin (10 mg/L), which needed a wide-spectrum antibiotic, antifungal and, antiviral treatment (meropenem, colistin, linezolid, tigecycline, voriconazole, acyclovir).

Due to the patient's septic status the stem cell mobilization was postponed. At present the patient is admitted to our bone marrow transplant (BMT) unit for stem cell

mobilization and harvesting with cyclophosphamide and granulocyte colony-stimulating factor (G-CSF).

The patient agreed to the publication of her data, and the institution where the patient had been admitted, approved the publication of the case.

DISCUSSION

In the case presented above we decided to stabilize the status of our patient by initiating chemotherapy treatment, followed by stem cell mobilization and harvesting. In this way the autologous stem cell transplantation will be performed after a previous treatment of the disease.

In cases similar to ours, a multidisciplinary approach is necessary. The possible presence of spontaneous fractures on pathological bone, due to secondary osteoporosis in some cases, needs prompt orthopedic treatment. On the other hand, renal disorders, frequently present in case of MM & PLC patients, need nephrologic treatment and hemodialysis.

PARTICULARITY OF THE CASE

One of the most important particularities of our case is the sudden and atypical onset of the PCL with the appearance of acute renal failure, with the necessity of hemodialysis.

The polyserositis revealed at diagnosis (bilateral pleurisy, minor pericarditis, average amount of ascites) was a medical challenge in establishing a correct and early diagnosis.

The concomitant presence of osteolytic lesions and spontaneous pathological bone fractures in association with acute renal failure should raise each doctor's suspicion of possible myelomatous pathology.

CONFLICT OF INTEREST

Nothing to declare.

REFERENCES

1. Albarracín F, Fonseca R. Plasma cell leukemia. *Blood Rev.* 2011;25:107-112.
2. Moreau P, San Miguel J, Ludwig H, et al. Multiple myeloma: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology.* 2013;24:vi133-vi137.
3. Fernández de Larrea C, Kyle RA, et al. Plasma cell leukemia: Consensus Statement on Diagnostic Requirements, Response Criteria, and Treatment Recommendations by the International Myeloma Working Group (IMWG). *Leukemia.* 2013;27:780-791.
4. Tiedemann RE, Gonzalez-Paz N, Kyle RA, et al. Genetic aberrations and survival in plasma cell leukemia. *Leukemia.* 2008;22:1044-1052.
5. Gertz MA, Buadi FK. Plasma cell leukemia. *Haematologica.* 2010;95:705-707.
6. Hanbali A, Alrajeh A, Rasheed W. Plasma cell leukemia mimicking hairy cell leukemia. *Hematol Oncol Stem Cell Ther.* 2015;8:91-92.
7. Van de Donk NWCJ, Lokhorst HM, Anderson KC, Richardson PG. How I treat plasma cell leukemia. *Blood.* 2012;120:2376-2389.
8. Jakab Sz, Lazar E, Benedek I Jr., et al. New Treatment Methods in Multiple Myeloma. *Journal of Interdisciplinary Medicine.* 2017;2:72-76.