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Case Report

IgA Plasma Cell Leukemia: A Rare Case

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ABSTRACT

Plasma cell leukemia (PCL) is a rare entity. It is classified as either primary PCL occurring at diagnosis or as secondary PCL in patients with multiple myeloma (MM).

The PCL is a rare disorder representing 1-2% of the diagnosed cases of MM. The median age is above 50 years. The electrophoresis shows a monoclonal protein in patients with PCL that is usually IgG (50%), IgA (15%), or in rare cases IgD or IgE (6%). Due to the rarity of this condition, only a few cases have been reported in the literature. It is characterized by its aggressiveness and poor prognosis. Through this case diagnosed in our laboratory we will describe the biologic particularities and prognosis of IgA primary PCL.

Keywords: Plasma cell leukemia-IgA-multiple myeloma

INTRODUCTION

Plasma cell leukemia (PCL) is a malignant proliferation of plasmocytic cells defined by a blood plasmocytosis greater than 2 G/L or a peripheral plasma cell count greater than 20% or an absolute plasma cell level greater than 2* 109/l [1].

It is a lymphoid proliferation that accounts for about 1 to 3% of acute leukemias. The primary form occurs de novo in a patient not followed for multiple myeloma (MM), and usually features a rapid clinical progression and short survival. The secondary form consists of the leukemic transformation of an already known MM. The clinical presentation of PCL approaches of acute leukemias more than MM. This aggressive presentation combines asthenia, bone pain, anemic syndrome and haemorrhages. There is a higher incidence of extra-medullary involvement especially hepatic (52%) and splenic (40%) [2]. the median survival is as low then six months.

CASE OBSERVATION

It's about a 53-year-old Moroccan patient, with no pathological history, admitted to emergency for asthenia, headache and vertigo evolving for 3 months. Extensive evaluation revealed a tumor syndrome made of hepatosplenomegaly.

A blood count showed hyperleucocytosis at 25 G/dl with anemia at 9 g/dl normochrome normocytic, thrombocytopenia at 60 G/µL, the blood smear showed 38% of the plasma cells. The myelogram showed a poor marrow with the presence of dystrophic plasma cells (Figure 1).

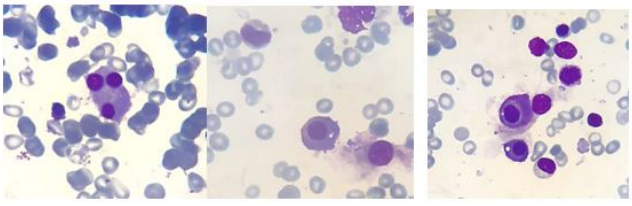

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Figure 1: Dystrophic plasma cells. (A) Multiple cores (B) Centralized core (C) Inflamed cytoplasm.

A complete biochemical assessment was realized and proved abnormalities for all the parameters processed. There was an inflammatory syndrome with a CRP elevated to 170 mg/L, a hypercalcemia to 110 mg/L, a kidney failure clearance of creatinine evaluated at 15 mL/min according to MDRD. The rate of the protides was 68 g/l. Serum protein electrophoresis makes it possible to highlight a peak of monoclonal appearance evaluated at 25 g/L migrating at beta 2 globulin (Figure 2).

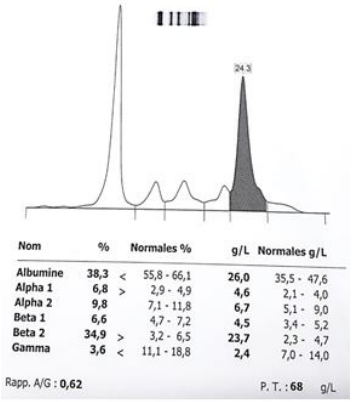

Figure 2: Serum electrophoresis results.

Immunofixation confirms the presence of an immunoglobulin monoclonal IgA type light chain kappa (Figure 3).

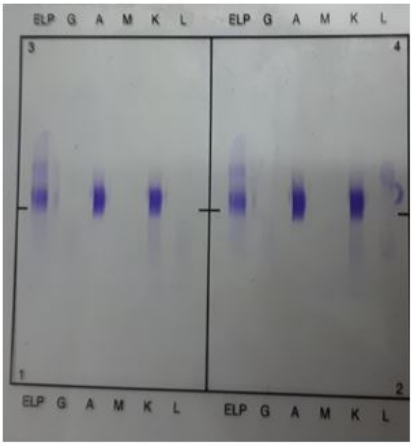

Figure 3: Immunofixation results.

Quantitative immunoglobulins performed by immune-turbidimetric method revealed an IgG of 4 g/dl IgA, 27 g/l, IgM, 0.2 g/l, Kappa, 9.53 g/l and Lambda, 0.52 g/l.

Free light chains showed kappa 194 mg/L, lambda 2.5 mg/L, and a markedly high kappa-lambda ratio 77.6.

Other abnormal laboratory studies were elevated LDH>500 IU/l; high beta 2 macroglobulin levels 4 mg/l.

Flow cytometry revealed the presence of CD19+, CD20+cell population expressing the CD117 and CD56 aberrant markers. Other markers like CD138 and CD38 were negative. Kappa light chain was also positive.

A radiologic bone series was negative.

Based on these biological findings our patient was diagnosed with IgA plasma cell leukemia.

The patient died from heart failure due to major hypercalcemia before the start on supportive care and chemotherapy.

DISCUSSION

The incidence of PCL is estimated between 2 and 4% in patients with multiple myeloma (MM) [2,3]. PCL results from a monoclonal proliferation of usually secreting plasma cells. The median age of onset is between 52 and 65 years, approximately 10 years younger than the median age observed in MM [2]. Although data are limited, it seems that, as with MM, LP is more common in African Americans than in Caucasians [3]

Clinically, PCL has a more aggressive presentation than MM, with an extensive involvement of visceral organs [4].

The diagnosis of PCL is essentially biological. It is based on data from the blood count and MGG-stained blood smear, which indicates the presence of a blood plasma cell count greater than 2 G/L or a circulating plasma cell count greater than 20% of the leukocyte count [1]. Compared to MM, PCL is more frequently responsible for anemia, thrombocytopenia [2].

Plasma cells are sometimes difficult to identify in blood smears and the use of immunophenotyping in ambiguous forms is essential for diagnosis [3]. PCL is also often characterized by light chain or non-secretory disease. It more frequently expresses CD20, CD44, CD45, CD19 and CD23, while CD27, CD56, CD71, CD117 and HLA-DR are more rarely detected [5] Notably, CD38, which is an important target for immunotherapy in MM, is constantly expressed in PCL [5].

It is also necessary to carry out a cytogenetic analysis by the fluorescent hybridization technique (FISH) of the clonal plasmocyte population. The presence of a translocation (4,14) involving the gene coding for the heavy chains of immunoglobulins or of a deletion of 13 is a factor of poor prognosis [6]

The assessment is completed by a myelogram or osteo-medullary biopsy to characterize the plasmocytes found [4]. These investigations must be completed by a comprehensive biochemical assessment. There is more frequently a renal failure and hypercalcemia in the LPC than in the MM, and very often in the LPs. This can partly be explained by a higher proportion of chain diseases slight [7].

An additional radiological procedure is requested. Of note, osteolytic bone lesions seem to be less frequent in PCL than in MM [5].

This patient had primary IgA plasma cell leukemia, which occurs in only about 15% of cases of plasma cell leukemia. In 50% of cases the serum monoclonal protein is IgG [8]. IgD paraprotein is uncommon in plasma cell leukemia; rarely, IgM and IgE have been reported [1]. Our patient had a markedly elevated IgA paraprotein, which may have increased the likelihood of developing hyperviscosity [9].

Patients have higher levels of β2 microglobulin and lactate dehydrogenase and lower levels of hemoglobin and albumin in comparison with MM [7].

From a prognostic point of view, the survival of patients with PCL is short. Historical median survival varies from 4 to 12.6 months with a 5-year survival rate of less than 10% in all studies [10].

CONCLUSION

Plasma cell leukemia is a rare disease with a poor prognosis. The diagnostic criteria of PCL are under discussion in the international myeloma community. It has characteristics that are common with MM but also has clinical, biological and prognostic features.

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