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PLASMA CELL LEUKEMIA: AN AGGRESSIVE DISEASE, A CAUSE OF DIAGNOSTIC ERROR

MOHAMED KEITA[1,2], SOKHNA AISSATOU TOURÉ[1,2,3], ELIMANE SEYDI BOUSSO[4], ALIOUNE BADARA DIALLO[1,2], NATA DIENG[4], SERIGNE MOURTALLA GUEYE[1,2], BLAISE FELIX FAYE[1,2,3], MOUSSA SECK[1,2,3], SALIOU DIOP[1,2,3]

[1]National Blood Transfusion Center, Dakar, Senegal

[2]Department of Clinical Haematology, CNTS, Dakar, Senegal

[3]Department of Haematology, Cheikh Anta Diop University, Dakar, Senegal

[4]Department of Clinical Haematology, HPD, Senegal

ABSTRACT

Plasma cell leukaemia (LP) is a rare and aggressive plasma cell tumour that manifests as significant clonal expansion of plasma cells in the bone marrow and peripheral blood. It is considered primary when it presents at initial diagnosis and secondary when it occurs in patients with pre-existing multiple myeloma (MM). Because of the high frequency of extramedullary injuries, plasma cell leukaemia is a major source of diagnostic error that can lead to fatality. We report a case of plasma cell leukaemia diagnosed at the clinical haematology department of the CNTS in Dakar (Senegal).

KEYWORDS: Plasma cell leukaemia, prognosis, diagnosis.

INTRODUCTION

Plasma cell leukaemia (LP) is a rare and aggressive plasma cell tumour that manifests as significant clonal expansion of plasma cells in the bone marrow and peripheral blood [(1)]. It is considered primary when it presents at the initial diagnosis and secondary when it occurs in patients with pre-existing multiple myeloma (MM) [(2)]. It is characterized by intrinsic genomic instability, high proliferative activity, and the coexistence of multiple adverse clinical and biological features, resulting in poorer outcomes compared to multiple myeloma (3). Because of the high frequency of extramedullary injuries, plasma cell leukaemia is a major source of diagnostic error. We present a rare case of a patient diagnosed with primary plasma cell leukaemia (PLL) who presented with atypical signs that progressed acutely to multiple organ failure.

Observation

The woman was 52 years old without particular medical history, referred to the clinical haematology department of the National Blood Transfusion Centre in Dakar for the exploration of a bicytopenia. The onset of her symptoms dates back to about a month before she was admitted to our ward, marked by the onset of uncontrollable vomiting associated with abdominal pain. Faced with these manifestations, she first consulted a gastroenterology department where she received an abdominal ultrasound which showed a homogeneous hepatomegaly without any detectable solid or liquid lesions. Biological investigations ruled out an infectious liver disease. However, the complete blood count showed bicytopenia consisting of anemia at 10 g/dL and thrombocytopenia at 120 G/L. In view of these results, she received symptomatic treatment and then was referred to a haematology department for the exploration of this bicytopenia. About a month later, she presented herself to the emergency room of our department (clinical haematology); On admission, she presented a worsening of her symptomatology with the occurrence of hematemesis of great abundance, non-febrile obnubilation with no sign of neurological localization. The entrance examination showed an anaemic syndrome poorly tolerated from a haemodynamic point of view (a regular tachycardia at 125 beats per minute, a diffuse systolic murmur at all focal points of cardiac auscultation of functional appearance), a haemorrhagic syndrome (hematemesis of great abundance, disseminated ecc hymotic purpuras), acute respiratory distress with saturation of 89% in ambient air and hepatic overflow at 3 cm at the level of the right midclavicular line. There was no lymphadenopathy or splenomegaly on examination; other vital signs at baseline were normal. Faced with this emergency table, the complete blood count showed aregenerative normochromic anemia at 5.3 g/dL associated with thrombocytopenia at 21 g/L while the leukocyte count was within normal limits. The activated partial thrombosis time was extended to 45 seconds uncorrected by the addition of a control plasma. The biologist had reported pan agglutination and plasma hyperviscosity during the procedures for blood grouping and viral B serology. These comments led to a systematic performance of other biological explorations, in this case serum protein electrophoresis, which showed a peak of 75.2 g/l in the monoclonal gamma globulin zone. Immunofixation of serum proteins revealed the presence of Kappa-like IgG. Cytological examinations had objectified the presence of dysmorphic plasma cells at the medullary level (69%) and peripheral plasma cells at 46%: these cytological aspects were compatible with plasma cell leukemia (46% circulating plasma cells) ***(Figure 1)***.

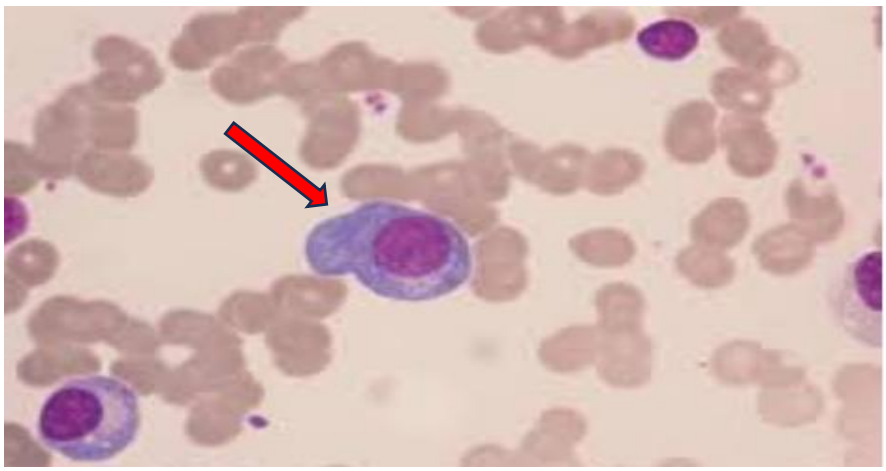

Figure 1 *(Dysmorphic plasma cells on blood smear (red arrow).)*

The prognostic score of the ISS allowed him to be classified as stage III (beta2microglobulinemia at 5.9 mg/l). In short, the diagnosis was plasma cell leukemia with systemic involvement (hepatic, renal, neurological and probably pulmonary). ***Table I***

Complete blood count		**Standards**
White Blood Cells (G/L)	8,63	4 – 10
Polynuclear neutrophils (G/L)	3,72	1,8 – 7
Lymphocytes (g/L)	1,96	1,5 – 4
Monocytes (g/L)	2,88	0,1 – 0,9
Hemoglobin (g/dl)	5,3	12 – 14
VGM (fl)	83,2	80 – 90
TCMH (pg)	27,7	27 – 32
CCMH (g/dl)	33,3	32 – 36
Platelets (G/L)	21	200 – 400
Reticulocyte count (G/L)	77	80 – 120
Hemostasis assessment		**Standards**
Prothrombin level (%)	75	70 – 100
TCA Sick / TCA Control	1,6	≤ 1.2
Mixed TCA (Patient + Control) / Control TCA	1,5	≤ 1.2
Biochemistry		**Standards**
LDH (IU/L)	654	120 – 246
Beta2 microglobulinemia (mg/l)	5,9	0,97 - 2,64
Aspartate transaminase (UI/L)	81	6 - 25
Alanine transaminase (IU/L)	12	6 - 25
Total bilirubin (mg/l)	21,8	< 10
Direct bilirubin (mg/l)	11,7	< 3
Creatinine (mg/L)	61	7 - 12
Glomerular Filtration Rate (CKD-EPI) (ml/min)	8,42	95 (± 20)
Natremia (mmol/l)	131,2	135 - 145
Kaliemia (mmol/l)	3,22	3,5 – 5,3
Albuminemia (g/l)	15	> 30
Corrected serum calcium (mg/l)	125	88 - 104

Inflammatory assessment		Standards
Sedimentation rate (mm) at 1 hour	100	< 15 mm in the 1st hour
C-reactive protein (mg)	13,2	< 6 mg/l

Table I *(Laboratory Characteristics of the Patient.)*

summarizes the patient's laboratory characteristics. Therapeutically, she received transfusion support and oxygen therapy without improvement of his clinical picture. Two hours after her admission to intensive care, she died in a state of cardiovascular shock.

DISCUSSION

Plasma cell leukemia is a rare and highly aggressive plasma cell tumor that develops in 0.5-4% of patients with multiple myeloma (MM). As with MM, LP is more common in African Americans and Black Africans than in Caucasians [4]. Kyle RA et al found a median age between 52 and 65 years old in a study of 869 cases [5]. Another epidemiological study involving 291 patients diagnosed between 1973 and 2004 found a median of 67 years [6]. It is therefore a condition that occurs in young subjects, unlike multiple myeloma, which is a pathology of mature adults. Our patient was diagnosed at the age of 52 with probably primary plasma cell leukemia; There was no history of multiple myeloma. The clinical picture is more aggressive than that of MM with a greater frequency of extramedullary lesions present in 23 to 100% of cases depending on the series, the most important are hepatic and splenic lesions found in 52% and 40% of primary LP cases respectively [7]. Our patient had already consulted for vomiting of a trivial appearance for which she received symptomatic treatment before being received in our department a month later for hematemesis. On physical examination, she presented with hepatomegaly, bone marrow failure syndrome and obtundiness. Other patients may present with pleural effusion, neurological deficits due to CNS involvement, or palpable extramedullary soft tissue plasmacytomas [8].

The diagnosis of LP is biological, based on blood count data and MGG-stained blood smear that shows a circulating plasma cell count greater than 5% of the leukocyte count [2]. Plasma cells are sometimes difficult to identify on blood smears and the use of immunophenotyping in ambiguous forms is essential for diagnosis. The work-up is completed by a myelogram or bone marrow biopsy, serum protein electrophoresis with immunofixation and a biochemical assessment. Compared to MM, LP is more frequently responsible for anemia, thrombocytopenia, hypercalcemia, renal failure, and higher serum levels of LDH and ß2-microglobulin (a reflection of tumor mass) due to the aggressive course of the disease, including a higher tumor burden and a higher proliferation index [9]. Due to the delay in the specialized consultation, our patient presented a sudden deterioration of her clinical picture with a multi-organ involvement (renal, pulmonary, neurological and probably pulmonary) which was life-threatening.

CONCLUSION

The study of this case allowed us to show that plasma cell leukemia is an aggressive disease that can have various circumstances of discovery. Larger-scale studies with good clinical-biological descriptions

are needed in our countries in order to be able to diagnose this pathology at an early stage and manage it optimally.

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