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Plasma Cell Leukemia: About Two Cases

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Abstract**—** *Our study reports two cases of secondary plasma cell leukemia to unknown and undiagnosed multiple myeloma; a female patient to plasma cell by the presence of 29% of plasma cells at the peripheral level, and a female patient B R, 52 years old, presented with anemic syndrome, metrorrhagia and diffuse bone pain. In terms of blood count, bicytopenia with severe anemia and thrombocytopenia at 11G/l, the blood smear noted blood plasmacytosis at 21%, suggesting plasma cell leukemia. Both patients benefited from other biological, radiological and other specific examinations to complete the diagnosis. In both patients, the clinic was polymorphous and aggressive with a progression towards death for the patient X, and the patient B R was put under a VTD protocol with a noticed evolution. The purpose of this study comes, on the one hand, from the rarity and aggressiveness of plasma cell leukemia, and on the other hand, it comes from the diagnostic and therapeutic difficulties with an uncontrolled and vague evolution. This should lead practitioners to diagnose, treat and monitor multiple myeloma, and to seriously think about it by standing against any blood plasmacytosis*.

Keywords**—** *Immunophenotyping, plasma cell leukemia, myelogram, plasma cells*.

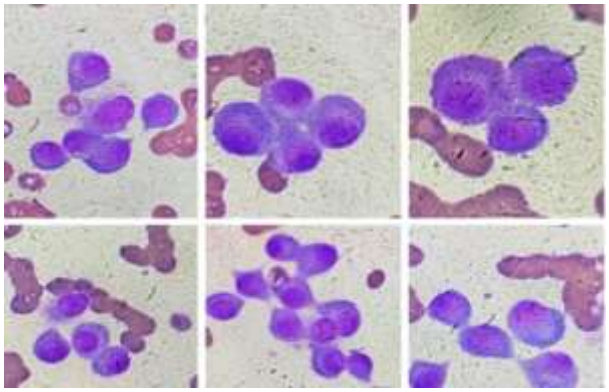
I. INTRODUCTION

Plasma cell leukemia is a specific and rare entity within multiple myeloma. In the past, it was defined by a blood plasma count ≥ 2G/L of leukocytes or greater than 20% of the blood count [1] and recently only with a count >=5% of leukocytes. . In most cases, it is secondary to progressive multiple myeloma, with an estimated frequency of 40-50%. [2, 3] Primary plasma cell leukemia is a very rare entity, characterized by its aggressiveness and poor prognosis, with an estimated frequency of 1-4% [1, 4]. Unlike multiple myeloma, it is mainly found in young people. Clinical expression is polymorphous, associating a tumor syndrome with signs of bone marrow failure as in other forms of leukemia [3, 4]. Current treatment is not well codified. In our study, we report two cases, the first to be described in our laboratory department at the My Ali Echerif regional hospital in Errachidia, to show the diagnostic, therapeutic and evolutionary difficulties of this type of pathology, and to consider it in the presence of revealing signs and, in the slightest doubt, to carry out immunophenotyping for a rapid treatment due to its seriousness.

II. COMMENTS:

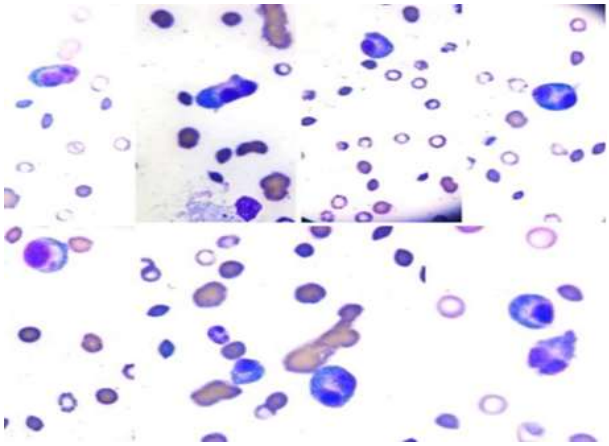
Case 1:

Patient X, aged 55, admitted to the emergency department with asthenia, pale skin and mucous membranes, cervical adenopathy, headache, infectious syndrome with respiratory signs, no hemorrhagic syndrome and no tumour syndrome. The symptomatology had been evolving for 2 months with the appearance of painless cervical adenopathy, and antibiotic therapy was instituted, combining C3G, quinolone and imidazole for 10 days. The clinical course was unremarkable, but the adenopathies persisted and increased in volume, associated with a continuous fever and subjective weight loss. The pneumopathy became severe and she was referred for intensive care. The blood count showed a hyperleukocytosis consisting of WBCs at 19 G/L, PNNs at 2.1G/L, lymphocytes at 2.3G/L, plasma cells at 5.51G/L; anemia at 6.8g/L and a platelet count of 189G/L. The blood count was checked by smear, showing significant anisopoikilocytosis with numerous red blood cells in rolls, and a plasma cell count of 29%.



Picture 1: Plasma cells on blood smear stained with MGG obj 100

The myelogram showed a rich marrow of increased density, with a scarcity of megakaryocytes, and 35% medullary plasmacytosis.



Picture 2: Medullary smear stained with MGG obj 100

Serum protein electrophoresis showed a monoclonal peak at the gamma level, while serum protein immunofixation was not

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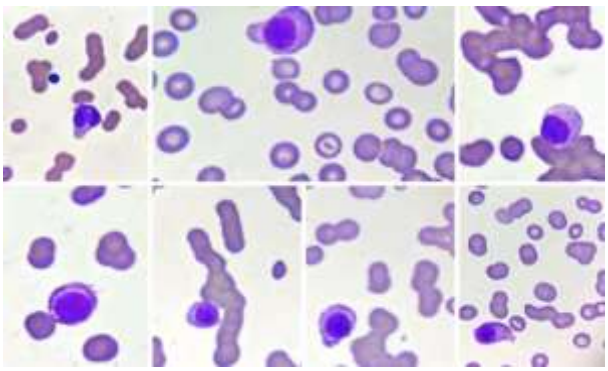
performed due to a lack of resources. X-rays of the skull, pelvis and dorsolumbar region revealed no lytic lesions. The rest of the biological work-up was carried out: CRP 51mg/l, blood culture negative. Bilirubin assay showed BIT at 21 mg/l and BIC at 10 mg/l. Symptoms persisted despite treatment and haematological resuscitation, with deterioration in general condition, the appearance of more profound bicytopenia with severe anaemia at 4g/dl and severe thrombocytopenia at 12G/L. We therefore decided to carry out immunophenotyping on the marrow. The patient died a week later with probable cerebral haemorrhage, and the immunophenotyping results suggested a diagnosis of plasma cell leukemia secondary to unknown multiple myeloma.

Case 2:

Mrs. BR, 52, married with 4 children, underwent surgery 20 days ago for a uterine fibroid. She presented to the emergency department with an anemic syndrome associated with a hemorrhagic syndrome consisting of metrorrhagia, infectious syndrome and diffuse bone pain, The internist prescribed a full biological work-up, radiological examinations and other measures to be monitored on the patient as well as to organize the management. The interrogation revealed the symptomatology asthenia, cutaneous-mucosal pallor evolving since four weeks, and the metrorrhagia persists since one week.

Biologically, the patient had thrombocytopenia of 50,000 on a previous CBC, and the new haemogram showed bicytopenia with severe normocytic normochromic aregenerative anaemia, hemoglobin 5.6g/dl, thrombocytopenia 11 G/L, WBC 3 G/L, PNN 0.7G/L, lymphocytes 1.9G/L and reticulocyte count 82,000/mm3.

MGG-stained blood smear showed red blood cells in rolls and 21% small plasma cells.

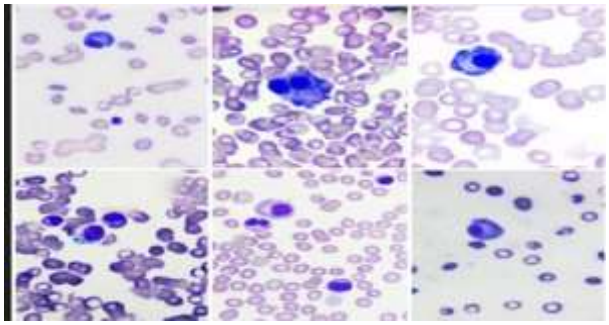


Picture 3: Blood smear stained with MGG obj 100

Biochemical tests showed plasma creatinine 87mg/l, urea 4 mg/l.

Liver function tests were normal, CRP 127mg/l and hypercalcemia 124mg/l. Bonce Jones proteinuria was very positive.

Myelogram showed increased marrow density with megakaryocytes, 32% bone marrow plasmacytosis with dystrophic, large, double- nucleated, small-nucleated plasma cells.



Picture 4: Medullary smear stained with MGG obj 100

Serum protein electrophoresis revealed a monoclonal peak in the Gamma zone, quantified at 41g/l.

Serum immunofixation confirmed this IgG Kappa monoclonal phenomenon. Immunophenotyping was consistent with plasma cell leukemia, with cells expressing CD19, CD20, CD38, CD45, and not expressing CD 138.

Radiologically, pelvic ultrasound revealed an enormous intrauterine hematoma. TAP CT images suggested peritonitis with lytic images, raising suspicion of multiple myeloma.

Nuclear magnetic resonance imaging revealed a plasmacytoma. On the basis of all these clinical-biological and radiological data, our diagnosis was plasma cell leukemia secondary to unknown and undiagnosed multiple myeloma.

The patient was transfused with 6 GCs, 55 PCs and 6 PFCs for DIC. She was on antibiotic therapy and progressed well, with a control CRP of 18 mg/L.

Hyperhydration with corticosteroid therapy to reduce hypercalcemia. She also benefited from two hemodialysis sessions to rebalance the distribution of water and other substances (urea, creatinine, etc.).

Background treatment was instituted, and the patient was put on a VTD protocol with good progression.

III. Discussion

Plasma cell leukemia is a rare lymphoproliferative disease characterized by the malignant proliferation of plasma cells in the bone marrow and peripheral blood. It is defined by the presence of more than 20% plasma cells, or a circulating plasma cell count greater than 2 G/L. The primary form (60% of cases) is seen again in patients with no previous evidence of multiple myeloma, whereas the secondary form (40% of cases) is the leukemic transformation of a previously known multiple myeloma, and in this case represents the ultimate evolution of the disease, which is generally relapsed or refractory (only 1% of multiple myelomas will evolve into secondary plasma cell leukemia). (3.4)

The diagnosis of plasma cell leukemia is essentially biological. It is based on blood count and MGG-stained blood smear data. The use of immunophenotyping for ambiguous forms is essential for diagnosis. Evaluation is completed by myelogram or bone and bone marrow biopsy. These investigations should be complemented by a full biochemical work-up [2]. Compared with MM, LP is more frequently responsible for extra-medullary localizations, anemia, thrombocytopenia, hypercalcemia, renal failure and higher serum levels of LDH and β2microglobulin (2).

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In all cases, this is a rare entity with a poor prognosis, especially as there is no known effective treatment.

IV. CONCLUSION

Plasma cell leukemia is a rare and serious hematological malignancy with a poor prognosis. We report two rare cases of plasma cell leukemia secondary to multiple myeloma, unknown and undiagnosed in our laboratory at My Ali Echerif regional hospital in Errachidia, in order to highlight the diagnostic, therapeutic and evolutionary difficulties of this type of pathology.

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