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(CASE REPORT)

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Plasma cell leukemia: A rare case

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Abstract

Plasma cell leukaemia is an extremely rare haematological malignancy with a poor prognosis.

It is defined by the presence in the circulating blood of a number of plasma cells greater than 2 giga/L or greater than 20% of the leukocytes. It presents in two forms: secondary plasma cell leukaemia complicating a known multiple myeloma, and primary plasma cell leukaemia, which is leukaemic in origin.

We report the observation of a 52-year-old Moroccan patient who presented with bone pain and general deterioration 3 months before hospitalisation.

The haemogram revealed normocytic normochromic anaemia at 8.1 g/dL and hyperleukocytosis. The blood smear showed the presence of 38% of plasma cells. The myelogram confirmed the diagnosis of APL by showing a rich marrow with 63% dystrophic plasma cells.

In this case, we emphasise the importance of careful cytological examination of the blood smear.

Keywords: Plasma cell leukemia; Blood test; Myelogram

1. Introduction

Plasma cell leukaemia (PCL) is the rarest but most aggressive variant of monoclonal gammopathies [1]. There are two forms: the primary form occurring de novo in patients without pre-existing multiple myeloma and diagnosed by immediate blood dissemination, and the secondary form, corresponding to a late event in patients with multiple myeloma.

We report a case of primary APL with a rapidly deteriorating course.

2. Case report

A 52-year-old Moroccan patient with no previous pathological history was admitted to the emergency department with inflammatory bone pain in the right upper limb, resistant to level 1 analgesics, which had been evolving for 3 months, in a context of altered general condition (weight loss of 10 kg, asthenia and night sweats).

On clinical examination, the patient was apyretic and presented with mucocutaneous pallor and pain on mobilisation and pressure of the right upper limb.

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There was no bleeding, infectious or tumour syndrome. The radiological workup showed lytic cookie-cutter images, not only of the right humerus but also of the skull and pelvis.

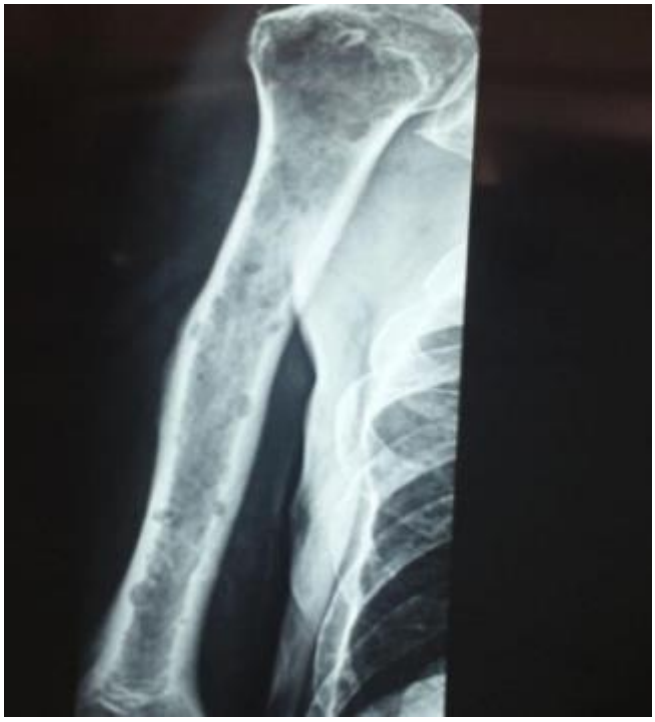

Figure 1 Standard X-ray showing die-cut geodes of the right humerus

The biological work-up revealed a normocytic normochromic anaemia with an aegerative count of 8.1 g/dL and thrombocytopenia of 100 giga/L associated with hyperleukocytosis of 16.2 giga/L. The blood smear showed a 38% circulating plasma cell count and rolling red blood cells (fig. 2).

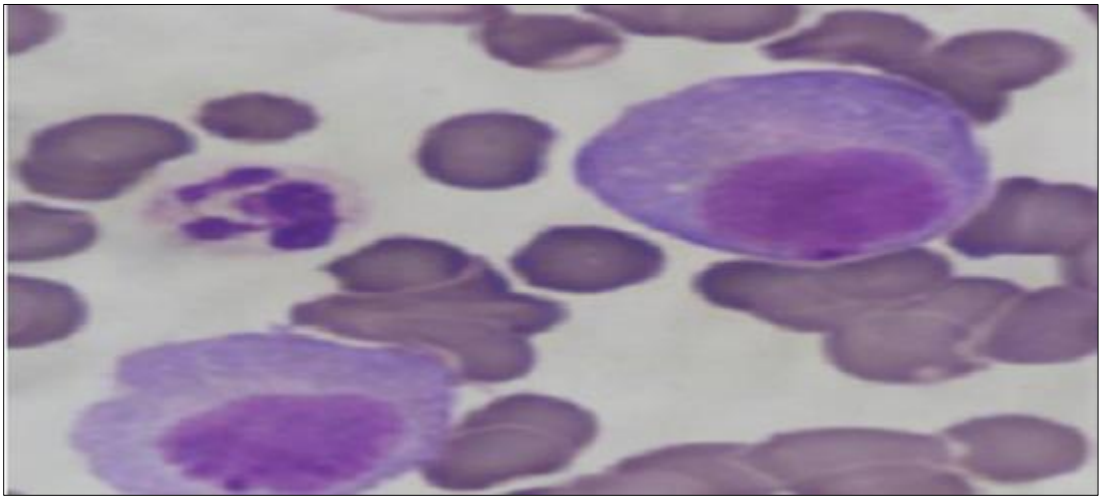

Figure 2 MGG-stained blood smear, magnification x 1000, showing circulating plasma cells

The myelogram showed a rich marrow, 63% of which was invaded by dystrophic plasma cells (fig.3).

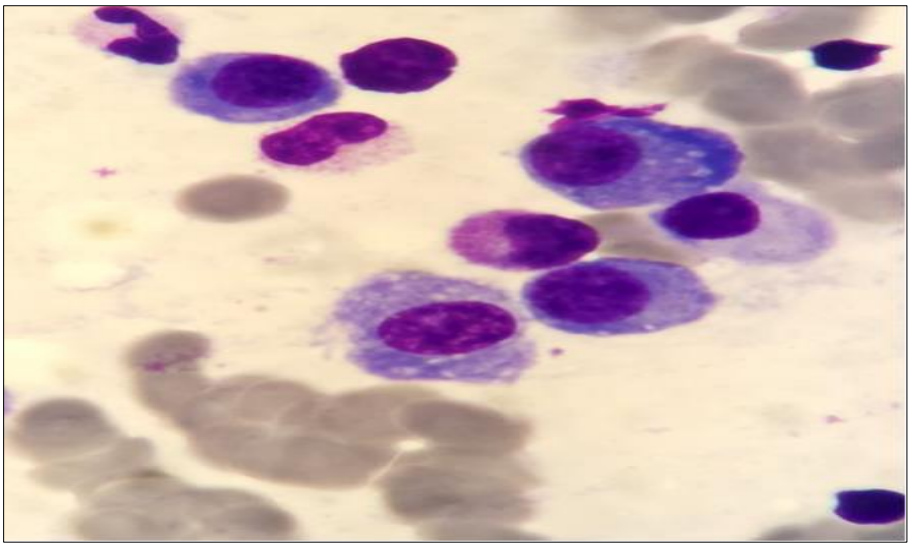

Figure 3 MGG-stained bone marrow smear, magnification x1000, showing bone marrow invasion by dystrophic plasma cells

In the biochemical work-up, there was an inflammatory syndrome with a CRP elevated to 170 mg/L, hyperproteinemia at 100 g/L, hypercalcemia at 120 mg/L, and renal insufficiency (creatinine clearance evaluated at 13 ml /mn according to MDRD).

Serum protein electrophoresis revealed an estimated 25 g/L monoclonal peak in beta2globulin.

Serum protein immunofixation confirmed the presence of a narrow band of high intensity at the level of IGA Kappa. The weighted immunoglobulin assay revealed an increase in IGA, 27 g/L and a collapse of the other classes including IGG 4g/L, IGM, 0.2 g/L, Kappa, 9.53 g/L and Lambda, 0.52 g/L.

The serum free light chain assay was as follows: kappa 194 mg/L, lambda 2.5 mg/L and a markedly elevated kappa/lambda ratio of 77.6.

Immunophenotyping showed the presence of CD19+, CD20+ cells expressing the aberrant markers CD117 and CD56. Kappa light chain was also positive.

In summary, we report a case of primary plasma leukaemia with IGA kappa. This is an unusual type of plasma cell leukaemia. The patient died before the start of chemotherapy.

3. Discussion

Plasma cell leukaemia was first described by Gluzinski and Reichenstein over a century ago. It is a rare form that accounts for only 2-4% of patients with multiple myeloma and 0.9% of all acute leukaemias [1]. It results from monoclonal proliferation of plasma cells and is defined by the presence of plasma cells in the circulating blood with a level of more than 2 giga/L or more than 20% of the leukocytes [1]. If there is any doubt, further investigations should rule out reactive causes such as infection, autoimmune disease or neoplasia. The median age of onset is between 52 and 65 years (about 10 years younger than the median age for multiple myeloma) and the sex ratio is 1 [3]. Our patient fits perfectly into this range.

The clinical presentation is more aggressive than that of multiple myeloma, with asthenia, bone pain, anaemia and haemorrhage. There is a greater frequency of extramedullary involvement (especially liver (52%) and spleen (40%) [4]).

Biologically, the frequency and severity of anaemia and thrombocytopenia appear to be greater in APL than in MM. With regard to monoclonal immunoglobulins, IGA forms are less frequent than in patients with multiple myeloma. In primary APL, there is a high percentage of light chain forms, which explains the frequency of renal involvement. Cytologically, plasma cells are classically well differentiated. In our patient, however, they were dystrophic. Hypercalcaemia, renal

failure and elevated serum β2 microglobulin are more common and our observation is one of them. On flow cryometry, plasma cells in APL share with multiple myeloma an expression of CD38 and CD138 surface antigens [5].

In contrast to multiple myeloma, overexpression of CD54 on myeloma cells facilitates their extramedullary migration and promotes tumour dissemination. Also, high expression of chemokine receptors (CCR1, CCR2 and CXCR4) is involved in the development of plasma cell leukaemia [7]. In addition, there is hyper expression of CD20 antigens and low expression of CD9, CD117, CD56 and HLA-DR antigens [6]. Secondary plasma cell leukaemia is characterised by the acquisition of CD28, which is responsible for increased cell proliferation and disease progression [6]. Finally, this immunological examination will allow the diagnosis to be established in the case of exceptional plasma cells with cytoplasmic projections mimicking tricholeukocytes [8, 9].Cytogenetics and molecular biology make it possible to identify numerous abnormalities, none of which is really specific.

Prognostically, APL is one of the poorest prognostic haemopathies. The historical median survival (before 2000) ranged from 4 to 12.6 months with a 5-year survival rate of less than 10% in many series. With the advent of new therapies and the increasing use of autologous haematopoietic stem cell transplantation, median survival has improved significantly (recently over 3 years). Treatment of APL is an emergency and should be initiated promptly. Currently, protocols based on proteasome inhibitors are recommended during the induction phase and allow rapid cytoreduction. This result should be consolidated by intensified therapy with autologous or allogeneic stem cell support in eligible subjects.

4. Conclusion

LAP is a very serious but fortunately extremely rare condition. The diagnosis is simple based on the discovery of a circulating plasmacytosis on the blood smear by a trained biologist.

Despite encouraging therapeutic results, especially for protocols including new molecules, there is still a long way to go to better understand the pathophysiological mechanisms and characterisation of the disease.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare no conflict of interests.

Statement of ethical approval

The authors appropriate statement of ethical approval.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

References

[1] Alghasham N, Alnounou R, Alzahrani H, Alsharif F. Plasma cell leukemia: Clinicopathologic, immunophenotypic and cytogenetic characteristics of 4 cases. Hematol Oncol Stem Cell Ther. 2015;8:71-7 pubmed publisher

[2] Bouaouad M. Thesis of medicine, N°: 113, 2011, faculty of medicine and pharmacy of Rabat, Morocco: 50.

[3] Costello R, Sainty D, Bouabdallah R, Fermand J, Delmer A, Divine M, et al. Primary plasma cell leukaemia: a report of 18 cases. Leuk Res. 2001;25:103-7 pubmed

[4] Ravinet A, Bay J, Tournilhac O. [Plasma cell leukemia]. Bull Cancer. 2014;101:1048-58 pubmed publisher

[5] Takahashi T, Kanno S, Itoh H, Yoshimoto M, Tsujisaki M, Fujino M. Two cases of nonsecretory multiple myeloma presenting as primary plasma cell leukemia. Intern Med. 2014;53:2121-5 pubmed

[6] Guieze R, Moreau A.S,Dupire S, Coiteux V,Facon T et al. Leucémie à plasmocytes, Hématologie, 2005, volume 11, numéro 3: 217-25.

[7] Oliveira A, Maria D, Metzger M, Linardi C, Giorgi R, Moura F, et al. Thalidomide treatment down-regulates SDF-1alpha and CXCR4 expression in multiple myeloma patients. Leuk Res. 2009;33:970-3 pubmed publisher

[8] Hanbali A, Alrajeh A, Rasheed W. Plasma cell leukemia mimicking hairy cell leukemia. Hematol Oncol Stem Cell Ther. 2015;8:91-2 pubmed publisher

[9] Majhi U, Murhekar K, Sundersingh S, Rajalekshmi KR. Primary cell leukemia with unusual presentation, Indian J hematol Blood Transfus, 2014, 30 (suppl1): 390-3.

[10] Chang H, Sloan S, Li D, Patterson B. Genomic aberrations in plasma cell leukemia shown by interphase fluorescence in situ hybridization. Cancer Genet Cytogenet. 2005;156:150-3 pubmedISSN : 2334-1009