A rare case of myeloma light chains: Data and discussion

Imane Tlamçani¹, Soukaina Oudrhiri Benaaddach², Kawthar Armani¹, Fatima El Boukhrissi², Mohamed El Baaj³, Kamal Doghmi², Mohamed Mikdame⁴, Mohamed Moudden Karim⁵, Moncef Amrani Hassanii², Lhoussine Balouch², and Youssef Bamou²

¹Hematology Laboratory, Central Laboratory of Medical Analysis, CHU Hassan II, Fes, Morocco
²Biochemistry Laboratory, Military Hospital Moulay Ismail, Meknes, Morocco
³Department of Internal Medicine, Military Hospital Moulay Ismail, Meknes, Morocco
⁴Department of Clinical Hematology, Military Hospital Mohamed V, Rabat, Morocco

ABSTRACT: Multiple myeloma is a little-known disease in young adults, it is rarely reported before the age of 30, even less if it is light chains. We report a case illustrating one of these exceptional situations. It has characteristics as organic sounding extended and favorable response to treatment despite diagnostic delay. We wanted to take the opportunity to raise awareness about the possibility of myeloma in adults at any age.

KEYWORDS: multiple myeloma; light chains; the young adult; organic sounding; diagnostic delay.

1 INTRODUCTION

The multiple myeloma, rare pathology at the young age, is defined by the malignant proliferation of a plasma cells clone being accompanied by the secretion of a monoclonal immunoglobulin [1]. The myelomas with intact immunoglobulin are most frequent (80%), those with light chains account for only 15 to 20% of the cases [2]. But they generate important complications and particularly at the renal level. Those can be avoided or slowed down if the disease is labelled in times. Where from importance of the early diagnosis, and there the place of biology is considerable. Indeed, while limiting itself to the clinic and to epidemiologic data, one can miss it with all the consequences which ensue from it. To enrich more the already available data , we considered useful to bring back and to discuss the case of a myeloma with light chains occurring before the age of thirty years: situation far from being common.

2 OBSERVATION

H.M. 29 years old woman was hospitalized for exploration of a paraparesy of the lower limbs. The interrogation and the exploitation of some old medical documents allowed to note the following datas: - no notable pathological histories, no tobacco nor of contact with pesticides, - seven months before: chronic inflammatory pains becoming invalidating at the sternocostal and dorsolumbar level, - microcytic hypochromic non-regenerative anemia uncorrected by an iron therapy, - deterioration of the general state.

The first intention physical examination revealed only neurological disturbances. A specialized examination ended in a paraparesy with thermo-algic hypoesthesy arriving up to the xiphoidian level, as well as an abolition of the cutaneous abdominal reflex. In the biological exploration, the complete blood count showed a bicytopenia: platelets in 130.000 elements/mm³ and a normochromic normocytic non-regenerative anemia (haemoglobin = 7 g/dl). The erythrocyte sedimentation rate (ESR) was in 48 mm. The imaging explorations, in particular by magnetic resonance imaging (MRI) showed
an osteolytic vertebral body collapse of D4, D7 and D10 with a spinal cord compression in the opposite side, and many lytic lesions on the coasts level and skull’s bones. The tumoral etiology with suspicion of bone metastasis was evoked.

A serum proteins electrophoresis (SPE) was performed at the same time (on Sebia’s Capillaries®), and revealed a small monoclonal peak in gamma globulins (10.9 g/l) with a serum protein rate in 75g/l (Figure 1).

Fig. 1. Serum proteins electrophoresis showing a small monoclonal spike in gamma globulin

The serum’s immunofixation on Agarose gel (Hydragel 1 IF®, Sebia) with the immuns serums anti-Ig G, anti-Ig A, anti-Ig M, anti-Ig E, anti-Ig D, anti-Kappa and anti-Lambda highlighted an isolated strip from light chains Lambda (Figure 2 and the gel with anti D and anti E: a and b). A Bence Jonce protein, made of free light chains, was also located at the urinary and serum level by immunofixation (Agarose gel hydragel I BJ®) (Figure2: c and d). The quantitative examinations showed [Kappa] = 5.2 mg/L and [Lambda] =103.3 mg/L. From where a ratio K/L = 0.05 (reference value: 0.26-1.65).
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**Fig. 2.** a et b : Serum immunofixation electrophoresis with agarose gel (Hydragel 1 YEW ®, Sebia) with immun sera anti-Ig G, anti-Ig A, anti-Ig M, anti-Ig E, anti-Ig D, anti-kappa and anti-lambda highlighting an isolated band of Lambda light chains.

c et d : Serum and urinary immunofixation electrophoresis with agarose gel (Hydragel 1 BJ ®) revealing the presence of a Bence Jones protein, made by Lambda free light chain.

A myelogram from a bone marrow sternal aspiration concluded to a dystrophic plasma cells infiltration up to 12% (Figure 3). The diagnosis of multiple myeloma with Lambda light chains was then made according to the Southwest Oncology Group (SWOG) diagnostic criteria. The value of the B2-microglobulin (9.05mg/l) classifies it at the stage III of International Staging System (the ISS). Other elements made possible to list it stage IIIA of Durie and Salmon (bone lesions, haemoglobin in 7 g/dl and a creatinin < 20 mg/l). The cytogenetics do not show anomalies: normal medullary chromosomic chart and the fluorescent hybridization in situ (FISH) also (no deletion p53 nor T (4; 14)).

**Fig. 3.** Medullary smear seen with the zoom x100 showing a dystrophic plasma cells infiltration of the bone marrow.
The patient was sent to the Mohamed V’s instruction military hospital for an appropriate care. After five cures of VTD (Velcade®= Bortezomib, Talidomide, Dexamethasone) followed by a therapeutic intensification (Melphalan 200mg/m2) and then an autograft, the patient is in complete remission with a eight month backward.

3 Discussion

The multiple myeloma (MM) is a malignant blood disease characterized by a clonal proliferation of tumoral plasma cells invading hematopoietic bone marrow [1],[2]. It represents 1.4% of all cancers and approximately 10% of hemato logic ones, what places it at the second rank in term of frequency after the lymphomas [3]. It affects preferentially men (sex-ratio H/F close to 1.4). Before the age of 40 years, this pathology is scarce, not exceeding 2% of the cases. It is even rarer before age of 30 years and only some cases were reported in the literature [1],[2]. This scarcity associated with an important organic repercussion constitutes one of the reasons which encouraged us to bring back this observation.

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The immunofixation blood test is the test of choice to characterize the type of the immunoglobulin (Ig) in cause [3]. Usually, it is practiced after having an electrophoretic aspect in touch with the myeloma, but sometimes even with a normal aspect of pace. Indeed, an electrophoresis without apparent anomalies does not exclude a immunoglobulinopathy (migration in β zone : IgA and light chains for example), a suspect clinical symptoms for example would be enough to require a immunofixation. The most met monoclonal immunoglobulins are IgG type (60% to 70% of the cases), followed by IgA (25% of the cases) whereas the light chains are only seen in 15% of the cases and Kappa is twice more involved than Lambda [2],[4]. The immunofixation enabled us to highlight in the blood and the urines of this patient light chains Lambda. Even then we register additional element in support of the rarity of this case (myeloma with light chains Lambda twice less frequent than with light chains Kappa).

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The patient in spite of her stage of infringement by the myeloma is currently in complete remission after having benefited from the therapeutic protocol reserved to the young people. Indeed, after an induction phase made of tritherapy with VTD and an intensification by the melphalan, the autograft was a success, the patient is until our days asymptomatic and all the parameters normalized and particularly the ratio K/L.
4 Conclusion

The multiple myeloma ends by complications which carry the patient, the early diagnosis allows a care in times. This way, while protecting the comfort of the existence of the patient, we manage to also improve his life expectancy. This pathology is very rare before the age of 30 years old, however this epidemiological data should not make us forget to think of it even by default. This is more justified if we consider the pejorative consequences of a diagnosis carried lately. An anemia with less than 10 g/dl of hemoglobin associated to an erythrocyte sedimentation rate even moderately high must bring us to prescribe at least a serum protein electrophoresis, even when the patient is less than 30 years old.

References


