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IgD-Lambda Multiple Myeloma without Renal Failure: An Unusual Presentation

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Abstract **Case Report**

Immunoglobulin D (IgD) multiple myeloma is a rare form of myeloma having often an aggressive course, affecting 2% of all cases. It shows frequentely renal failure, Bence Jones proteinuria and the difficulties of diagnosis. We report a case of a 59 years old man with IgD multiple myeloma associated with good renal function. The patient was hospitalized in the hematology department for bone pain and alteration of the general condition. A further evaluation for multiple myeloma revealed presence of a monoclonal protein band in Beta2 region in serum protein electrophoresis (SPE). Urine protein electrophoresis (UPE) showed presence of BenceJones proteinuria.The immunofixation electrophoresis (IE) of the serum reported as IgD-Lambda paraproteinaemia. Laboratory studies showed also an anemia , hypercalcemia, but fortunatuly, our patient did not have renal failure, as is common with myeloma. Bone marrow examination showed infiltration by plasma cells. Thus, the patient was put on chemotherapy and stem cell autotransplantation with a good clinical and biological improvement. This case is reported to emphasize the importance of performing IgD immunofixation routinely for all suspected multiple myeloma patients because many cases are erroneously diagnosed as light chain disease. And it describes a rare case of an even rarer and uncommon situation and highlights the fortunate situation of this patient's unfortunate disease.

Keywords: Immunoglobulin D multiple myeloma- Immunofixation- renal failure- overall survival.

INTRODUCTION

Multiple myeloma is a mature B lymphoid neoplasia defined as an acquired hematologic malignancy, characterized by clonal proliferation of plasma cells with complete or incomplete monoclonal immunoglobulin synthesis. It represents 1 % of all cancers, and 10% of haematological cancers [1].

IgG, IgA and light chain myeloma are the most widespread and account for 54%, 21% and 16% of all myelomas, respectively. Immunoglobulin D multiple myeloma is a rare form of myeloma having often an aggressive course, affecting 1% or 2% of all cases. It shows frequentely renal failure, Bence Jones proteinuria, amyloidosis, poorer prognosis, and difficulties of diagnosis than other multiple myeloma subtypes [2].

This report describes a rare case of an unusual condition and outlines the fortunate aspects of this patient's unfortunate diagnosis.

CASE REPORT

A 55 years old man, without any background history was admitted in an hospital center of onco-hematology for dorsolumbar and pelvic bone pain of progressive onset, associated with an altered general state. Physical examination revealed a spinal and bone syndromes, and mucocutaneous pallor. There was no lymphadenopathy or hepatosplenomegaly.

His complete blood count revealed normochromic normocytic aregenerative anemia (haemoglobin 9.7 g/dL) with normal white blood cell and platelets counts. Sedimentation rate was elevated to 80 mm in first hour.

Bloodchemistries showed: normal levels of urea (0, 36 g/L) and creatinine (9,7 mg/L), normal levels of serum total protein and albumin :79g/l and 38g/l. Beta2 microglobulin was normal : 3g/L. Albumin adjusted calcium was high: 112 mg/l. 24-hour proteinuria was 0.9 g/l. The serum protein

electrophoresis (SPE) revealed monoclonal band in beta2globulin region estimated at 5g/l associated with decrease in gamma globulins. Urine protein electrophoresis (UPE) showed presence of Bence-Jones proteinuria. However, the routine immunofixation electrophoresis (IE) of the serum and urine samples showed lambda light chains but was negative for anti IgG, A and M. Further immunofixation with IgD and IgE antisera identified IgD-lambda paraproteinaemia.

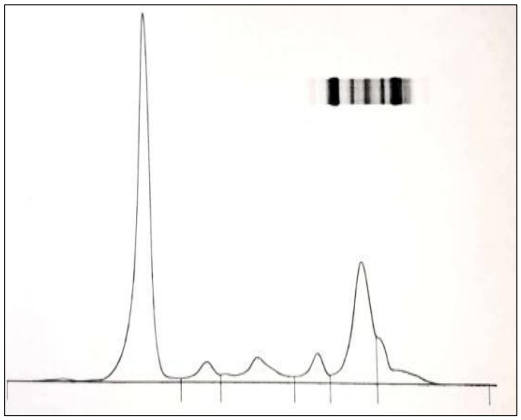

Figure 1: SPE showing a peak monoclonal in beta2globuline region+ decrease in gammaglobulins

Cranial profile radiograph demonstrated lacunar lesions with diffuse demineralization. Magnetic resonance imaging of the spine showed diffuse vertebral involvement.

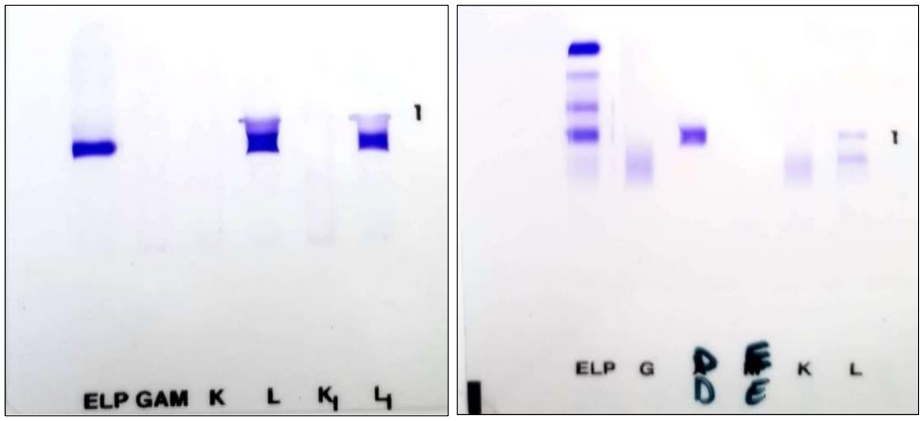

Figure 2: IE of the serum is negative for IgG, IgA and IgM and positive for IgD-Lambda paraproteine

The bone marrow aspirate confirmed a multiple myeloma with 79% of plasma cells mostly dystrophic: large size, intracytoplasmic vacuoles. Thus, The patient was put on protocol 6 BDT (bortézomib,dexamethasone, thalidomide) with biphosphonates.

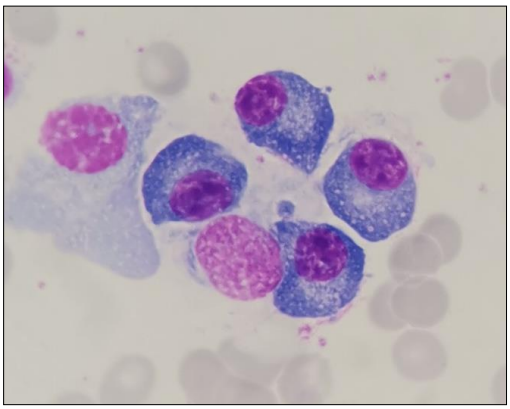

Figure 3: Bone marrow aspiration showing plasma cells (MGGx1000)

The evolution was marked by a complete hematological remission. The complete blood count was normal, the control myelogram showed 1% of plasma cells. The renal function and the calcemia count were normal, with a regression of the monoclonal peak on plasma protein electrophoresis.

The patient benefited from an stem cell autotransplantation with a very good improvement over 2 years follow-up.

DISCUSSION

Immunoglobulin D multiple myeloma is unfrequent, with an incidence of about 2% of all patients diagnosed with myeloma. Since the serum concentration of physiological IgD is much reduced compared to IgG and IgA, the M peak on serum protein electrophoresis (SPE) is very often quite small or even indistinguishable in IgD myeloma, which makes its diagnosis very challenging [3].

It tends to present at a younger age, favor male gender and be of more advanced stage [4].

Fatigue, weakness, pallor, and bone pain are the most common initial manifestations. However, In contrast to other myelomas, IgD myeloma has been shown to have more features of high-risk disease especially renal injury, Bence Jones proteinuria and hypercalcemia that were found in 20-40% of patients at diagnosis. Its explanation may be due to either cast light chain nephropathy or direct toxicity from intracellular crystals or the delay in diagnosis [5].

Laboratory analysis of IgD multiple myeloma cases by SPE often demonstrates a monoclonal peak in the beta, gamma or beta-gamma region. A large percentage of cases may have hypogammaglobulinemia or a normal serum electrophoretic profile. Bence Jones proteinuria is positive in almost all patients. Since IgD immunofixation is not routinely performed, many cases are misdiagnosed as non-secreting or light chain multiple myeloma [6].

Unlike other myelomas, lambda light chain predominance is a characteristic feature of IgD myeloma and is seen in 70% to 90% of cases which is the case for our patient [2]. This might be attributed to some preferential rearrangements of the heavy chain genes with the light chains [7].

Staging of multiple myeloma is based on the results of blood tests and imaging studies. Results of other tests may also help determine the stage, depending on the system used. Typically, 2 systems are employed to establish the stage of multiple myeloma: International Staging System (ISS) and Durie and Salmon classification. According to the ISS[8], the stage of the disease is I (4.3% of cases) if the beta 2-microglobulin level is less than 3.5 mg/l and the albumin level is equal to or greater than 35 g/l. Stage III (61.2% of cases) includes those with beta 2-microglobulin at 5.5 mg/l. Patients who did not meet the criteria for either of the other stages were classified as stage II (24.5% of cases). The median overall survival is 62, 44 and 29 months for stages I to III respectively.

Our case is exceptional owing to the absence of renal injury at presentation and the early disease of the patient at presentation, an uncommon beneficent status in IgD myeloma.

The treatment of IgD MM is similar to that of other subtypes of MM. Bortezomib and Thalidomide regimens are the most frequently used new therapies, they have shown excellent efficacy. The use of high-dose consolidation chemotherapy with new agents as well as autologous stem cell transplantation, has shown improved overall survival in IgD MM patients over the past 10 years [5].

CONCLUSION

IgD multiple myeloma without renal failure is a very rare disease affecting younger population with poor prognosis. It often poses a difficulty in differential diagnosis with non-secreting and light chain multiple myeloma, hence the interest in performing immunofixation with IgD and IgE immunserum. Early diagnosis and therapeutic management can avoid several complications, in particular renal failure, which is a frequent occurrence in IgD multiple myeloma and where patients end up on hemodialysis despite better control of the hematologic component.

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