http://jmscr.igmpublication.org/home/ ISSN (e)-2347-176x ISSN (p) 2455-0450 crossref DOI: https://dx.doi.org/10.18535/jmscr/v10i9.13



A Rare Case of Plasma Cell Dyscrasia – Kappa Light Chain Myeloma

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Abstract

We report anextremely rare case of Kappa light chain myeloma in a 70 year old female who presented with pancytopenia. Light chain myeloma is characterized by the inability of the malignant plasma cells to produce heavy chains, resulting in the exclusive production of light chains. Light chain myeloma tends to have a poor outlook compared to other myelomas since it's frequently more aggressive and often presents with Renal failure. Worldwide incidence of Light chain myeloma is approximately 15-20% among multiple myeloma patients. The incidence of lambda light chain myeloma is predominant among light chain myelomas.

Keywords: Plasma cell dyscrasia, Kappa light chain, Light chain myeloma.

Introduction

Light-chain multiple myeloma (LCMM) is a less frequent type of multiple myeloma (MM), with a more aggressive course and poorer prognosis. It is characterized by the inability of the malignant plasma cells to produce heavy chains, resulting in the exclusive production of light chains. ^{1,2} Therefore, no M-spike is visible in serum protein electrophoresis. Light chain myeloma tends to more frequently cause Renal failure, bone disease, and a buildup of light chain cells in organs (light chain amyloidosis). Worldwide incidence of approximately 15-20% among multiple myeloma patients.³

Case Report

A 70 -year-old female, with a history of diabetes mellitus, presented with easy fatiguability for a period of 2 months and weight loss for 3 months. On examination patient was normotensive with a

blood pressure of 126/87 mm Hg, heart rate of 70 bpm. Pallor was present, no lymphadenopathy. Per abdominal examination revealed hepatosplenomegaly. Other system examination was unremarkable. Routine investigations showed pancytopenia. Hemoglobin of 9.6 g/dl, WBC of 800 cells/cu.mm (ON SMEAR)Differential countcounted upto only 10 cellsout of which neutrophil 2/10, lymphocytes 7/10, and platelet count of 119000/ul. ESR 0f 150mm in the 1st hour, Serum LDH 400IU/L. Renal and Liver function test were within the normal limits. Corrected reticulocyte count was 0.1%, Peripheral smear microcytic hypochromic anaemia, leucopenia with lymphocyte predominanace and thrombocytopenia. In suspicion of hypocellular marrow, Bone marrow biopsy was done which showed 15% plasma cells. Bone marrow aspirate showed Atypical plasma hypercellular with erythroid cells, marrow hyperplasia and plasmacytosis suggestive of plasma

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cell dyscrasia. In view of multiple myeloma further investgations were done: serum calcium 7.9 g/dl, urea 17 mg/dl, creatinine 1mg/dl, Urine bence Jones proteins were Negative .X- ray skull and pelvis shows no lytic lesions. Serum electrophoresis was done for the patient andthere was no M-B and seen. No abnormal Band were detected in Urine electrophoresis also.

Further investigation for serum free light chains was done. It showed increase in serum free kappa light chain (7.67 g/L) as well as increase in serum free lambda light chain (3.51 g/L). Serum Free kappa lambda light chain Ratio was elevated (2.18). Patient was diagnosis as Kappa light chain myeloma.

Patient was not a transplant candidate due to physiologic age >70 years. Studies have proven that Continuous use of the lenalidomide dexamethasone combination appears to be superior to the MPT (melphalan, prednisone, thalidomide) regimen, making it a standard of care for older adults with myeloma. Hense the patient was started on dexamethasone 40 mg Q week and Oral Lenalidomide 25 mg daily × 21 days q 4 week. Patient was planned for cytogenetic studies, but patient was expired 1 month after initiation of therapy due to neutropenic sepsis. Light chain myeloma caries a poor prognosis compared with other subsets of myeloma.

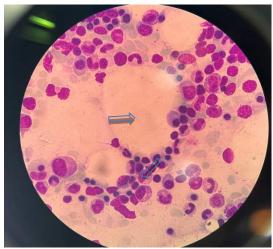


Figure 1 Light microscopic view of bone marrow biopsy showing plasma cells

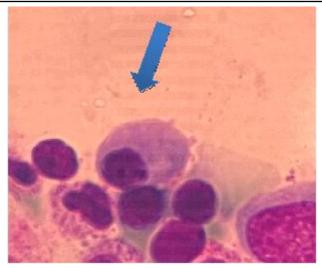


Figure 2. The cell marked is a plasma cell which has an eccenteric round nucleus with peri nuclear halos. Presence of >10% plasma cells confirm the diagnosis of Plasma cell dyscrasia.⁷

Serum protein Capillary electrophoresis

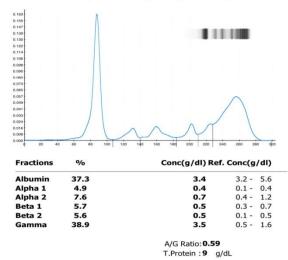


Figure 3. Serum electrophoresis of the patient showing NO M- Spike /M BAND, Increase in Total Protein, Polyclonal rise in gamma globulin, low albumin globulin ratio.



Fig 4: Xray skull lateral view (LEFT) and Pelvis AP view (Right) showing no lytic lesion.

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Discussion

Although most multiple myeloma (MM) cases are characterized by the detection of a monoclonal immunoglobulin in the serum, about 15% of the patients present only with immunoglobulin light chains, detected either in the urine or serum or both. 4,5 These patients are designated as having lightchain (LC) MM. A blood or urine test will show an increased level of free light chains, and more importantly, an abnormal ratio of kappa to lambda light chains. Patients with light chain myeloma have light-chain deposition in many organs, including the kidneys, liver, and heart, as well as in the skin and nervous system. Proteinuria or renal insufficiencies are the most common presenting complaints. Pancytopenia can be rarely seen in the later stages of myeloma once malignant plasma cells that take up space in the healthy bone marrow, eventually crowding out and impairing the production of normal white blood cells, red blood cells and platelets. Unfortunately, the prognosis for myeloma patients with acute renal failure is poor despite aggressive intervention.⁶ Light chain myeloma tends to have a poor outlook compared to other myelomas since it's frequently more aggressive and often presents with kidney failure. Serum beta 2microglobulin is the single most powerful predictor of survival and can substitute for staging. Other factors that may influence prognosis are the presence cytogenetic abnormalities hypodiploidy which can be evaluated by karyotype, fluorescent in situ hybridization (FISH). Continuous use of the lenalidomide and dexamethasone combination appears to be superior to the MPT (melphalan, prednisone, thalidomide) regimen, making it a standard of care for older adults with myeloma.

However, our patient presented with atypical presentation of pancytopenia, no lytic bony lesions and no renal involvement with bone marrow plasma cells of 15% and elevated serum Kappa light chains making it an extreme rare case of plasma cell dyscrasias.

Take Away Message

- Presence of >10% plasma cells in bone marrow confirm the diagnosis of Plasma cell dyscrasia.
- Light chain myeloma present only with immunoglobulin light chains, detected either in the urine or serum or both.
- No M-spike is visible in serum protein electrophoresis in light chain myeloma
- Renal insufficiencies and bone lesions are the most common presentation.
- Light chain myeloma has a more aggressive course and poorer prognosis.

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