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General Medicine

NON-SECRETORY MYELOMA PRESENTING AS PANCYTOPENIA : A CASE REPORT

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ABSTRACT Multiple Myeloma(MM) is a malignancy of haematopoietic cells characterized by clonal proliferation of plasma cells.Nonsecretory myeloma(NSM) is a rare variant of multiple myeloma(MM) accounting for 1-5% cases.The clinical features and radiological findings are similar to MM but the hallmark is absence of monoclonal immunoglobulins in serum and/or urine electrophoresis.Most cases present with anaemia,bone pain and infections.We report a case of 75 year old man who presented with pancytopenia and was subsequently diagnosed as non-secretory myeloma.

KEYWORDS : multiple myeloma, non-secretory, pancytopenia

INTRODUCTION

Multiple myeloma is a haematological malignancy characterized by clonal proliferation of plasma cells. There is production of large quantities of defective immunoglobulins. Nonsecretory myeloma(NSM) is a rare variant of MM accounting for 1-5% casesThe clinical features and radiological findings are similar to MM but the hallmark is absence of monoclonal immunoglobulins in serum and/or urine. Most cases present with anaemia, bone pain and infections NSM presents a diagnostic challenge for clinicians . Our patient presented wiyh pancytopenia hence we report this case.

CASE REPORT

A 75 year old Indian male was admitted to our hospital with complain of low back pain and fever for last 2 months. Fever was low grade and not associated with chills or rigorBack pain was dull aching in character, continuous in nature and was relieved by analgesics. . He also complained of generalized weakness and easy fatiguability. He was earlier treated at a private clinic where 3 units of whole blood and 2 units of platelet was transfused. There was no significant past medical history nor any history of similar illness in family.

On examination patient was conscious, oriented and febrile. He had pallor but no icterus or oedema. His pulse rate was 108 beats/minute and BP was 110/70 mm Hg. On auscultation bilateral vescicular breath sounds were present. CVS examination revealed tachycardia. Abdomen was soft with no organomegaly.

Investgations showed a haemoglobin of- 5. 4 g/dl with MCV-90fl (Macrocytic & Normochromic), total leucocyte count was 1200 with Neutrophils-25%, Lymphocytes-65%, Monocytes-7%, Eosinophils-2%, Basophils-1%. Platelet count was 90000/ cubicmililitre. Peripheral blood smear showed pancytopenia with no target cells, schistocytes or blast cells. His blood urea was 53mg/dl, serum creatinine was 1. 3gm/dl. Serum electrolytes revealed a serum calcium-7. 4gm/ dl. ESR was 160(0-20). Vitamin b12, folic acid and iron profile was normal. So a bone marrow biopsy was planned to evaluate pancytopenia. Bone marrow examination revealed Plasma cell level of>10% and suggestive of Plasma cell dyscrasia. An evaluation for MM was started. In light of patients complain of chronic backache a computed tomography of L-S spine was done which showed multiple round osteolytic lesions of vertebral bodies and iliac bones(Fig 1&2). Serum and urine protein electrophoresis (SPE&UPE) was conspicuous by absence of monoclonal spike(M-protein). Since absence of M-protein can also occur in oligo-secretors so a free light chain assay was done which was normal. Serum immunoglobin test showed a raised serum IgG&IgE level with normal serum IgA level. Beta2microglobulin was 4mg/dl and serum albumin was 3. 1mg/dl. Based on clinical findings, bone marrow biopsy and radiological profile a diagnosis of Nonsecretory myeloma(NSM) was made. Patient was transfused whole blood and plateles and was subsequently referred to a higher haematological centre for further management.

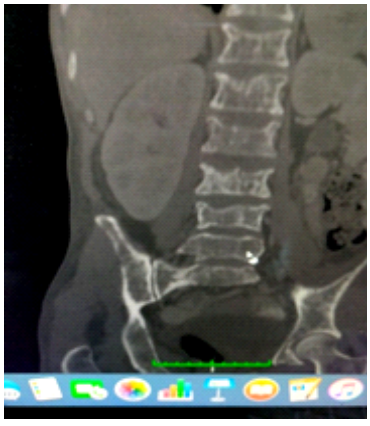

Fig1:Contrast enhanced ct of L-S spine showing multiple round lytic lesion of vertebral bodies and iliac bones

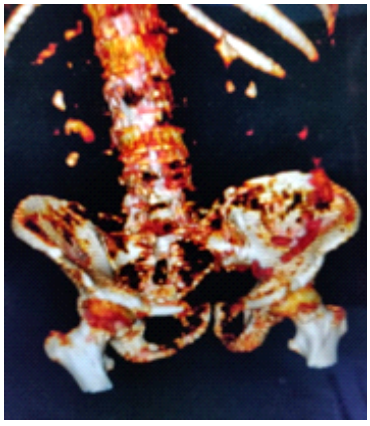

Fig 2:Reconstruction image showing round lytic lesions.

DISCUSSION

Multiple myeloma(MM) is a malignancy of haematopoietic stem cells characterized by clonal proliferation of plasma cells. These plasma cells produce immunoglobulins(Ig) which are non-functional. Nonsecretory myeloma (NSM) is a rare variant of multiple myeloma accounting for 1-5% cases. It is charecterised by absence of detectable immunoglobulins in serum and/or urine. The diagnostic criteria of NSM includes:

1) No M protein in serum and/or urine with immunofixation.
2) Bone marrow clonal plasmacytosis>10%
3) Myeloma related end organ damage(CRAB)

- a)Hypercalcemia
- b)Renal dysfunction

- c)Anaemia
- d)Bone lesions

Recent advancement in serum free light chain assay has enabled further classification of NSM into subtypes.

a) Oligosecretors-Produce only free light chains which can be detected by serum free light chain assay.
b) Non-producers-Do not produce any Ig(Light chainor heavy chain)
c) True nonsecretors-Produce Ig molecules but do not secrete them.
d) False nonsecretors-These are MM variants that have detectable intracellular Igs but no measurable extracellular component. Intracellular Igs

The clinical findings are similar to that of MM with most patients presenting with bone pain, anaemia and infections. The diagnostic workup for NSM includes SPEP, UPEP, Serum free light chain assay and imaging. Modalities include x-ray/CT/PET scans. CT and PET are useful in identifying the site of bone disease and also differentiate between active and quiescent lesions at completion of treatment as well as during follow upAll patients with suspected MM or NSM should have bone marrow biopsy to demonstrate plasmacytosis.

Once diagnosis of (NSM) is made the treatment protocol is similar to that of MM and consists of chemotherapy and/or autologous bone marrow transplant. The course, progression and prognosis of NSM is not well studied IN 1986, Smith et al. released a case series that included 13 NSM patients, in which NSM patients had a median survival of 46 months compared to 22 months for secretors. Moreovermonitoring response to treatment is difficult in NSM due to lack of detectable immunoglobulins(Ig) in serum and/or urine. Serial bone marrow biopsy can be considered the gold standard but invasiveness and cost make it practically less feasible. So methods that are used include obtaining a baseline CRAB criteria and then assess for any improvement after initiation of treatment. Repeat PET and CT are employed to monitor response.

CONCLUSION

Nonsecretory Myeloma is a rare variant of multiple myeloma and poses a diagnostic challenge for clinicians due to absence of Igs in serum and/or urine. Any patient suspected of MM should undergo bone marrow biopsy and serum free light chain assay to distinguish secretors from nonsecretors. The treatment protocol is similar to that of MM. The overall survival and prognosis is comparable to its secretory counterpart but further research is needed.

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