Case Report
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Non Secretory Multiple Myeloma Presenting as Pathological Fracture Bilateral Humerii: A case report

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Abstract

Non Secretory Multiple Myeloma (NSMM) is a rare variant of multiple myeloma which is characterised by the absence of monoclonal gamma-globulins in blood or urine. The patients usually present with multiple lytic lesions in bones with pain, anaemia, hypercalcemia, normal renal function, and positive clonal proliferation of plasma cells and atypical plasma cells in bone marrow biopsy. We present a case report of a 45 year old female with bilateral pathological fracture of humerus. Skeletal survey showed multiple lytic lesions throughout the body with absence of monoclonal gamma globulins on serum immunoelectrophoresis. Bilateral plating was done for humeral fractures and chemotherapy was instituted. Fractures united and patient's condition improved.

Keywords: Non Secretory Multiple Myeloma, Pathological Fracture, Management.

Introduction

Plasma cell neoplasms comprise of spectrum of diseases characterized by clonal proliferation and accumulation of terminally differentiated B cells. This spectrum comprise of conditions as benign as Monoclonal Gammopathy of Undetermined Significance (MGUS); indolent conditions like Waldenström's macroglobulinemia on the one end and more malignant disorders like multiple myeloma and plasma cell leukemia on the other. These disorders have a common feature on serum protein electrophoresis that is the production of single monoclonal peak (M component). Among patients with multiple myeloma, 70% have their M component comprising IgG and 20% having IgA. Production of monoclonal light chains only represents 5 - 10%. Less than 1% of patients produce monoclonal IgD, IgE, IgM or have non-secretory myeloma [1]. Non Secretory Multiple Myeloma (NSMM) comprise of 1-5% of all multiple myeloma cases [2]. It presents with diagnostic difficulty as there is absence of detectable monoclonal proteins in the serum as well as urine [1]. The plasma cells are unable to secrete the monoclonal immunoglobulin into the blood. Immunoglobulin is typically present in the cytoplasm of neoplastic plasma cells as confirmed by immunofluorescence study. We report this case due to it being very rare and presenting a diagnostic challenge to the treating surgeon.

Case Report

We hereby report a case of 45 years old female who came to us with the complaints of pain to right arm following trivial trauma. For this X-ray of right whole arm AP and lateral view was done which showed pathological fracture of right humerus mid 1/3rd through a lytic lesion in the bone (fig.1). This prompted us to investigate the cause of lesion. We did the skeletal survey of the patient which revealed multiple punched out lesion in the calvarial bone, also lytic lesions were seen in multiple dorsolumbar vertebrae, proximal femur, ilium, ischium and opposite humerus. Blood investigations were done which mainly revealed anaemia and thrombocytopenia

Laboratory results: Hg:9.6 g/dl WBC:7.6 x 103/uL , hematocrit:19.6% MCV: 91.6 fl MCH:27.5 pg, MCHC: 30.2 g/dL, PLT- 56 x 103/uL ,AST: 19 U/L,

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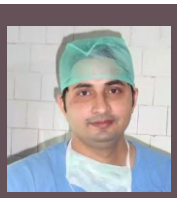
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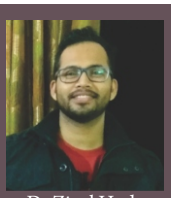
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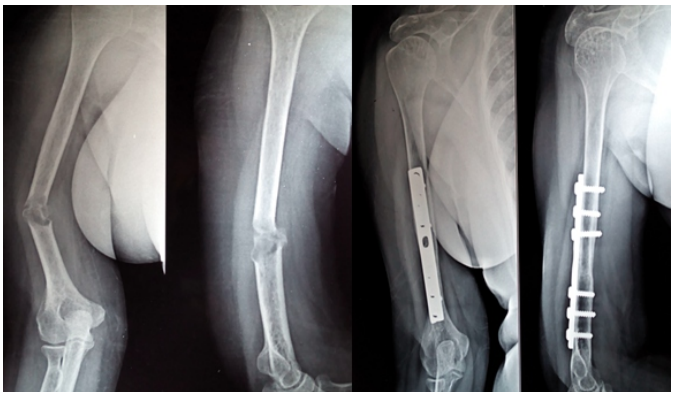

Figure 1: Shows pathological fracture of right humerus middle 1/3rd which was fixed with narrow DCP and the 1 year post operative check x-ray shows united fracture

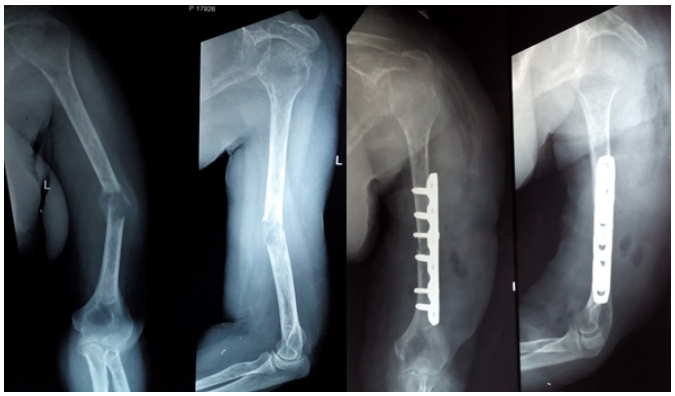

Figure2: Shows pathological fracture of left humerus middle 1/3rd which was fixed with narrow DCP and the 6 month post operative check x-ray showing uniting fracture

ALT:12 U/L, BUN:7.0 mg/dL, Creatinin: 0.9 mg/dL, Uric acid:5.6 mg/dL, Calcium: 3.05 mmol/L, Protein:6.40 g/dL, albumin: 3.91 g/dL globulin: 2.49 g/dL. MR imaging of dorsolumbar spine was done which showed anterior wedge collapse of D6 vertebra and decreased height of D9 and D10 vertebrae with altered signal intensity in the body and posterior element of vertebrae and bilateral ribs at multiple levels, sacrum, bilateral ilium and right humerus both showing heterogenous enhancement on post contrast sequence. Serum immunoelectrophoresis was done which showed no M spike, normal A:G ratio. Histopathological examination of the tru-cut biopsy from the lytic lesion showed sheets of plasma cells with interspersed capillaries which confirmed the diagnosis of non-secretory multiple myeloma. For the fracture of right humerus excision of lesion with open reduction and internal fixation with narrow DCP was done and the fracture united uneventfully. Two years later the patient again presented with pathological fracture of of left humerus for which she was again operated and lesion excision with open reduction and internal fixation with narrow DCP was done (Fig.2). The patient was put on Melphalan and prednisolone combination chemotherapy. Patient had subjective improvement and both fractures united in subsequent follow ups.

Discussion

NSMM primarily manifests as multiple lytic lesions of bone, anemia, hypercalcemia and thrombocytopenia. Mainly skull, ribs, humerus and sternum are involved [3]. The clinical manifestations of NSMM are similar to plasma cell myeloma, except the lower incidence of renal insufficiency and a better prognosis [4]. The diagnosis of non-secretory multiple myeloma is mainly made on the basis of multiple lytic bone lesion, anemia, hypercalemia, and a normal serum and urine protein electrophoresis and immuno-electrophoresis. At times it becomes difficult to make diagnosis of NSMM due to lower percentage of plasma cell in marrow and the bone marrow aspirate obtained from routine sites like sternum and hip often only showed a picture consistent with generalized medullary reaction against chronic infection or metastasis [5].

It is a subject of debate that why is there an absence of detectable monoclonal M-protein in patients with NSMM. Intracellular immunoglobulin can be detected in plasma cells implying defective immunoglobulin secretion or abnormal Ig structure which is not capable of being transmitted by the secretory pathway [6].

NSMM patients are treated the same way as those of secretors. Melphalan-Prednisolone combination chemotherapy remains a standard regimen for symptomatic patients over 70-year old and younger patients who are not candidate for transplantation. More

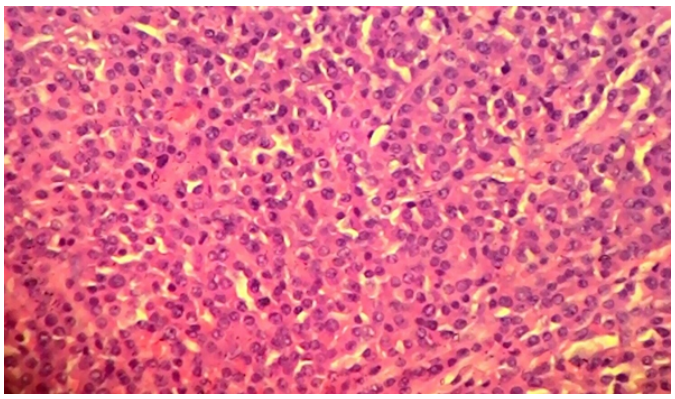

Figure3: Shows sheets of plasma cells with interspersed capillaries occassional plasmablasts and binucleate forms are also seen

aggressive chemotherapies do not show superiority over Melphalan. VAD regimen is less myelotoxic, can be used as initial treatment for myeloma patients who are candidates for transplantation. Biphosphonates are recommended for patients with bone lysis. Zolendronic acid 4 mg or Pamidronate 90 mg is given every 3-4 weeks. Clinical judgment regarding the palliative benefit of intravenous biphosphonate and the inconvenience of using biphos-phonate injection must be kept in mind. Intermittent evaluation for albuminuria and azotemia should be performed in patients on chronic biphosphonate treatment.

Fractures are generally believed to be common in patients with multiple myeloma as a result of lytic bone lesions, generalized bone loss, and/or elevated bone turnover from excessive cytokine production, but the actual risk of pathologic versus osteoporotic fractures has not been quantified. At diagnosis, pathologic fractures are the presenting feature in 30% of cases of Multiple Myeloma [7]. But risk of pathological fracture in patients with NSSM have not been quantified yet.

Conclusion

In the presence of multiple lytic lesion on skeletal survey, we should not exclude the diagnosis of multiple myeloma merely on the absence of immunoglobulins in blood or urine. We should confirm the diagnosis of multiple myeloma on bone marrow biopsy. We should keep the diagnosis of non secretory multiple myeloma in presence of lutic lesion and absence of M-spike on immunoelectrophoresis.

References

1. Riedel DA, Pottern LM. The epidemiology of multiple myeloma. Hematol Oncol Clin North Am 1992; 6: 225-30.
2. Grogan TM, Van Camp B, Kyle RA, Muller-Hermelink HK, Harris NL. Plasma Cell Neoplasma. Pathology & Genetics Tumours of Haematopoietic and Lymphoid Tissues. ES Jaffe, NL Harris, H Stein, JW Vardiman eds. WHO Classification of Tumor IARC Press, Lyon. 2001: 142-56.
3. Dave SS, Dunbar CE. Multiple myeloma. Handbook of Clinical Hematology. Rodgers DP, Young NS eds. Lippincott Williams & Wilkins Philadelphia, 2005.
4. Smith DB, Harris M, Gowland E, Chang J, Scarffe JH. Non-secretory multiple myeloma: a report of 13 cases with a review of the literature. Hematol Oncol. 1986 Oct-Dec;4(4):307-13.

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