INTRODUCTION

Plasma cell neoplasms represent a spectrum of diseases characterized by clonal proliferation and accumulation of terminally differentiated B cells. The spectrum includes conditions as benign as Monoclonal Gammopathy of Undetermined Significance (MGUS); indolent conditions like Waldenström’s macroglobulinemia and more malignant disorders like multiple myeloma and plasma cell leukemia. These disorders share a common feature of production of single monoclonal peak (M component) on serum protein electrophoresis. 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

Multiple myeloma is diagnosed on the basis of major and minor criteria comprising tissue diagnosis, monoclonal gammopathy, bone marrow plasmacytosis, lytic bone lesions, and suppressed uninvolved immunoglobulin.2

More recently, International Staging System (ISS) for multiple myeloma has been established on the basis of beta-2 microglobulin and serum albumin levels.3

We are presenting a case report of non-secretory myeloma, which is a rare variant in the spectrum of plasma cell neoplasms.

CASE REPORT

A 42-year-old male presented to our department with history of a lump at the upper part of sternum present for the last two years. The swelling was small in size to begin with but enlarged slowly over time. Patient complained of slight pain in the lump for about two months with generalized weakness. There was no associated history of fever, cough, headache, abdominal pain, or any other lump. On examination, there was a lump at the manubrium sternii; about 4 x 3 cm in size, mildly tender, immobile and firm in consistency. The baseline blood picture with differentials, renal and liver function tests and uric acid levels were essentially normal. Chest X-ray revealed multiple small lytic lesions involving especially both scapulae and clavicles. Ultrasound abdomen was normal except for mild fatty infiltration of liver. CT scan of the chest showed an expansile lytic lesion involving manubrium sternii with extraosseous extension. There were also lytic expansile lesions involving right 3rd, 4th, and left 6th ribs and the thoracic spine. The lump was excised, which turned out to be a plasmacytoma on histopathology. Bone marrow biopsy was normal with plasma cells representing only 4%. Serum protein electrophoresis was essentially normal. Quantitative immunoglobulins were then performed which showed IgG (8.42 g/dl), IgA (1.28 g/dl) and IgM (0.41 g/dl) to be within normal limits.

Urine test was negative for Bence Jones protein. Other relevant findings were raised levels of beta-2 microglobulin (82M, 3478 ng/ml) and low levels of serum albumin (2.6 g/dl). Serum calcium was markedly raised at 14.5 mg/dl.

X-ray skull revealed multiple punched out lesions of varying sizes. Final diagnosis of non-secretory myeloma was made as it fulfilled one major criterion of tissue diagnosis of plasmacytoma and one minor criterion of multiple lytic lesions in the skeleton.
The patient was put on oral Melphalan and Prednisolone along with monthly injection of Zometa. Subjective improvement in symptoms and serum $\beta^2$M was noted on follow-up.

DISCUSSION

Western literature reveals that the incidence of multiple myeloma is 01% of all malignancies in the white population and 02% in blacks. Among hematological malignancies, it constitutes 10% of the tumors, second in frequency to non-Hodgkin’s lymphoma. Non-secretory myeloma is a rare variant of multiple myeloma characterized by absence of detectable M protein in serum and urine. Multiple myeloma is diagnosed on the basis of major and minor criteria comprising tissue diagnosis, monoclonal gammopathy, bone marrow plasmacytosis, lytic bone lesions, and suppressed uninvolved immunoglobulins. This system has drawbacks in that some patients with disease-related symptoms might not meet the criteria for myeloma because of a low marrow involvement and/or Monoclonal (M)-protein. More recently, International Staging System (ISS) for multiple myeloma has been established on the two of most powerful prognostic factors in multiple myeloma i.e. beta2 microglobulin and serum albumin levels.

About 5 - 10% of symptomatic patients may have less than 10% of plasmacytosis, as was the case in this patient. The diagnostic dilemma may be resolved in such cases with tissue diagnosis, lytic bone lesions and raised serum levels of $\beta^2$M (in the absence of renal failure). The presenting features of non-secretory myeloma, response to therapy and survival are similar to those of patients with measurable M-proteins. The patient responded well to Melphalan and Prednisolone, along with bisphosphonate Zometa, with subjective improvement of his symptoms and lowering of serum levels of $\beta^2$M. In the absence of any clinically measurable disease and normal bone marrow biopsy along with no M-component, cytogenetic studies and sensitive serum free light chain assays might help in reaching the diagnosis. Previous studies regarding clinicopathological features and immunological patterns of multiple myeloma from Pakistan have not reported the non-secretory variant of myeloma.

REFERENCES