A RARE CASE REPORT OF PLASMA CELL NEOPLASM IN A YOUNG PATIENT

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Abstract: Multiple myeloma is a disease of the elderly, extremely rare below 30 years of age. This case of multiple myeloma was diagnosed in a young lady aged 30 yrs. She presented with anaemia and bone marrow plasmacytosis. No lytic lesions were present. Serum protein electrophoresis was negative. However immunofixation electrophoresis showed monoclonal kappa light chain

Keyword: Multiple myeloma, Young patients, M-protein and plasma cells

INTRODUCTION: Multiple myeloma, a malignant plasma cell disorder is a disease of the elderly with peak incidence between 60-70 years. Less than 2% of cases have been reported below the age of 40 years. It is extremely rare in patients <30 years. The presenting clinical and laboratory features are similar to those observed in patients of all ages who have myeloma. However a higher proportion of young patients are reported to have on light chain myeloma. These light chains are highly nephrotoxic which may lead to end stage renal disease.

CASE HISTORY:
A 30 year old lady was admitted with complaints of leg pain and back pain of one month duration. She had low grade fever on and off for 1 month. General examination revealed pallor and bilateral pitting pedal edema. There was mild hepatosplenomegaly. Examination of other systems were unremarkable.

Laboratory investigation revealed a Hemoglobin value of 7g/dl and haematocrit value as 25.14g/dl. Red cell indices and coagulation profile were within normal limits. Urine examination showed albuminuria (3+).

Peripheral smear examination revealed normocytic normochromic anaemia. Bone marrow aspiration study showed a hypercellular marrow, predominating cells being plasma cells and their precursors(figure1). These plasma cells were large cells with abundant basophilic cytoplasm and eccentrically placed nucleus with a perinuclear hoff (figure 2). Few binucleate forms were also seen and plasma cells with cytoplasmic vaculations (Mott cells) were seen (figure 3).

figure 1: Majority of the cells in this smear are plasma cells and their precursors

figure 2: Plasma cells(100 X) figure 3: Mott cell(100 X)
Few plasmablast were also seen in the smear which are larger cells with central nuclei high N/C ratio fine chromatin and prominent nucleoli and without perinuclear hoff.

Monoclonality of the plasma cells was confirmed by immunohistochemistry which showed positive for immunoglobulin light chain kappa (figure 7).

Bone marrow trephine biopsy showed hypercellular bone marrow (figure 5) with replacement of normal haematopoetic elements by plasma cells and their precursors (figure 6).

Radiologically no lytic lesions were made out. Serum calcium was within normal limits (10.2 mg/dl). Serum lactate dehydrogenase was markedly raised. Other biochemical parameters were normal.

Rheumatoid factor and Antistreptolysin-O titre were negative which excludes polyclonal hypergammaglobunemia. Serum protein level was low (5.5g/dl) with reversal of A:G ratio (1:1). Serum protein electrophoresis showed a normal pattern. 24-hours urine protein estimation value was 1.7 g/day (>1g/ dl). Urine protein electrophoresis showed a band in alpha 1and beta region. Serum immunofixation electrophoresis confirmed the presence of monoclonal M band which was found to be free kappa light chain. Serum beta2 microglobulin was increased (2.7 mg).

DISCUSSION: The diagnosis of multiple myeloma is rare below the age of 30 years. Among 3278 patients with multiple myeloma at Mayo clinic USA only 10 patients (0.3%) were younger than 30 years at diagnosis. In a study by National cancer institute only 17 patients were under 30 years among 3815 pateints with myeloma. (0.18%) Hewell et al have reported frequency of 1%.
In this 30 year old female with anaemia, bone marrow studies showed high degree of marrow replacement by plasma cells. This case is confirmed as plasma cell myeloma as it satisfies the criteria proposed by

INTERNATIONAL MYELOMA WORKING GROUP which includes

- M-Protein in serum or urine
- Plasma cells in bone marrow
- End organ tissue impairment (one or more of the following)
  - Hypercalcemia
  - Renal insufficiency
  - Anaemia
  - Lytic bone lesion

Although serum protein electrophoresis was normal, Immunofixation electrophoresis confirmed the presence of M-spike corresponding to kappa light chain. This is because M band is due to monoclonal Ig or Ig fragment which may be Ig G in 55%, Ig A in 25%, Ig D in 1%, Ig M in 1% and only free light chains in 20%. When the monoclonal M protein is free light chain, it is excreted as soon as it is secreted. So only low levels of M-protein was present in the serum which could be detected only by immunofixation electrophoresis. Most of the studies showed negative Bence Jones protein. Only a report by Blade et al reported the presence of Bence Jones protein in 5 out of 10 cases. In the present study, Bence Jones protein was positive. Multiple myeloma in the young has an atypical presentation and has increased median survival rate

CONCLUSION:
The presence of monoclonal plasma cell proliferation in the bone marrow in addition to the demonstration of demonstration of kappa light chain by immunofixation electrophoresis confirmed this case as light chain secreting plasma cell myeloma. This category of myeloma rarely occurs in patients less than 30 years of age.

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An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Pre and Para Clinical Sciences