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Ig D Myeloma Presenting As Cauda Equina Syndrome in a Young Patient –A Rare Presentation

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Abstract: Ig D myeloma is the rarest subtype of multiple myeloma and accounts for 2 % of all symptomatic myelomas. Majority of patients who present with IgD myeloma has lambda light chain and present with advanced disease stages and are associated with somatic hyper-mutation of IgV regions. The survival of patients with IgD MM is shorter in comparison to those with immunoglobulin G (IgG) myeloma but the outcomes are improving with the use of novel. Here we present the case of a 47 year old male with Ig D myeloma who presented with cauda equina syndrome. The patient was treated with bortezomib lenalidomide and dexamethasone chemotherapy.
Keywords: Ig D myeloma; cauda equina; prognosis.

INTRODUCTION

Ig D myeloma is extremely rare and accounts for less than 2 % of all symptomatic myelomas. It has a very poor prognosis and presents generally presents with an advanced disease. Here we present the case of young male who presented with cauda equina syndrome and was diagnosed as a case of Ig D myeloma.

CASE HISTORY

Forty-seven year old male was evaluated for backache radiating to lower limbs. There was no history of fever, decreased urine output, weakness of upper or lower limbs.

Evaluation showed the presence of anemia with raised ESR. MRI Spine was taken as he had severe backache and that showed bulge of posterior cortex of S1 vertebral body with compression of cauda equine roots (Figure-1).

His renal function tests were impaired (blood urea 133 and serum creatinine 6.9) and he had an albumin globulin reversal. Skeletal survey showed the presence of lytic lesions in ribs, vertebra, sternum, scapula and multiple other sites. Computed tomography of the chest showed the presence of multiple lytic bone lesions with soft tissue involving the posterior aspect of the left ninth rib. Serum IgG was 822mg/dl, IgA was less than 40mg/dl and IgM was less than 20 mg/dl. Free kappa was 33.5mg/dl and free lambda was 6385 mg/dl with a kappa lambda ratio of 0.005. Serum protein electrophoresis showed a major band in gammaglobulin region [M spike of 1.48 g/dl] and serum immunofixation electrophoresis showed the presence of band in lambda alone. Further immunofixation electrophoresis was done to look for IgD and IgE bands and it revealed a monoclonal band corresponding to IgD and Lambda (Figure-2). Bone marrow study showed the presence of 50% plasma cells. Beta2 microglobulin was 11.6mg/dl. Urine BJP was positive. Thus the diagnosis of multiple myeloma IgD lambda ISS - 3 was made and patient was started on chemotherapy with bortezomib and dexamethasone. External beam radiation was given to spine in view of painful spinal lesions. The patient had a good clinical biochemical response and improvement in renal functions and is improving and is planned for consolidation with autologous stem cell transplantation.

DISCUSSION

Multiple myeloma (MM) is a hematological malignancy characterized by clonal proliferation of plasma cells and is associated with end organ damage in form of anemia, renal failure, bone lesions and recurrent infections. The monoclonal plasma cells in multiple myeloma usually produce immunoglobulins and are usually of the IgG, IgA, or IgM, subtypes. IgD myeloma is extremely rare and there are reports that these patients may have an inferior outcome when compared to other patients. The majority of patients with IgD myeloma has lambda light chain and present with renal dysfunction and features of advanced disease. The survival of patients with IgD MM is shorter in comparison to those with immunoglobulin G

(IgG) MM but the outcomes are improving with the use of novel agents and autologous transplantation**.**

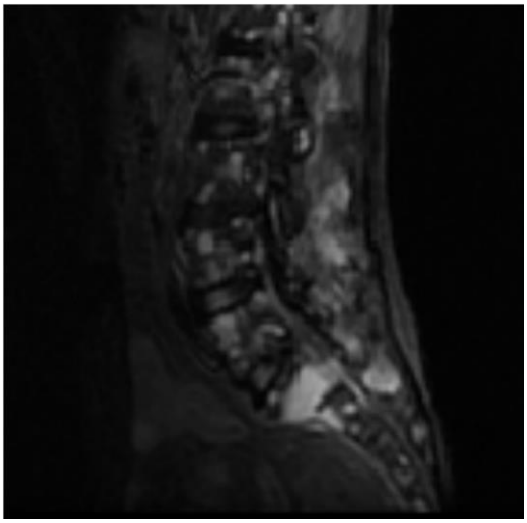

Fig-1: Contrast enhanced magnetic resonance imaging of the Spine showing bulge of posterior cortex of S1 vertebral body with compression of cauda equine roots

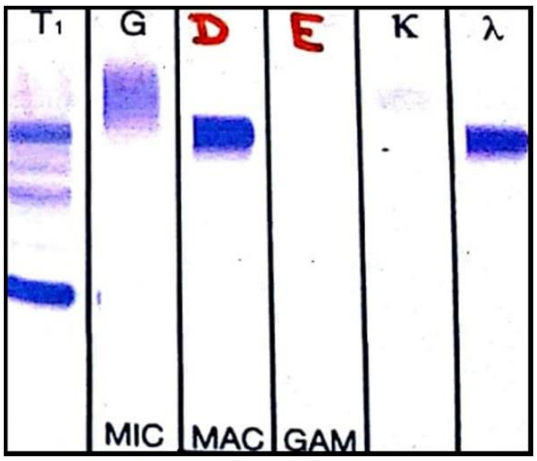

Fig-2: Immunofixation electrophoresis of the patient showing a monoclonal band corresponding to IgD and Lambda

IgD secreting plasma cells originate from germinal center B cells due to somatic hyper-mutation of IgV regions [1], the level of IgD in serum is 0 to 10 mg/dL. After IgD MM was first reported by Rowe and Fahey [2] in 1965, multiple studies have reported an IgD MM prevalence of approximately 1% to 2% of myeloma patients [3-7]. IgD MM occurs in relatively younger patients, with a median age of 50 to 60 years and predominantly occurs in males. The patients with IgD myeloma generally has a small or absent M-protein spike on electrophoresis, as well as extramedullary involvement, osteolytic lesions, presence of systemic amyloidosis, and renal failure [4-7]. Another feature of IgD MM is the presence of advanced disease stage at the time of diagnosis. Shimamoto *et al*., reviewed 165 Japanese patients with IgD MM classified according to the Durie-Salmon (DS) staging system.[8] They found 7% of patients to be DS stage I, 22% DS stage II, and 71% DS stage III. Similarly, a staging of 379 IgD patients in another study [6] reported 6%, 17%, and 77% of the patients in DS stages I, II, and III, respectively. However, two studies found no significant relationship between DS stage and survival outcomes in patients with IgD MM [8, 9].

The patients are worked up as in other patients with MM, although there may be higher incidence of amyloidosis, renal failure and bone lesions. The treatment in patients with IgD MM has been similar to other patients with MM. Due to rarity of this disease there is no specific guidelines for the management of this disease. The patients are usually started on a combination containing IMIDs and proteasome inhibitor along with steroids. The response to treatment is usually similar to other types of multiple myeloma with response rate of up to 70% in various reports. The general prognosis in patients with MM is variable with some reports suggesting an inferior outcome while others suggest a similar outcome to other myeloma groups [10].

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