CASE REPORT
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Rare Presentation of Multiple Myeloma: A Case Report

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ABSTRACT

Multiple myeloma (MM) is a malignant tumor due to neoplastic plasma cell proliferation derived from a single B-cell lineage. These cells produce monoclonal immunoglobulins, most commonly either immunoglobulin G (IgG) or immunoglobulin A (IgA). The peak incidence of MM is in the seventh decade, whereas, it is a rare entity in young patients. The clinical presentations are noted in MM patients like fever, fatigue, weight loss, paraesthesias, hypercalcaemia, hyperviscosity, renal failure, bone pain or pathological fractures, cutaneous lesions, etc. We are presenting a case of 55 years female patient with MM. This presented headache for four months and low back pain for two weeks. Headache is a rare symptom in MM. Increased level of β_2 microglobulin, serum creatinine, and plasmablastic type myeloma cells suggest poor prognosis of myeloma patients.

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INTRODUCTION

Plasma cell tumors are B-cell lymphoid neoplasm comprising of multiple myeloma (MM), solitary bone plasmacytoma and extramedullary plasmacytoma. Multiple myeloma is a malignant proliferation of plasma cells leading to monoclonal proliferation of paraprotein. Normal plasma cells are derived from B cells and produce immunoglobulins that contain heavy and light chains. Normal immunoglobulins are polyclonal, which means that a variety of heavy chains are produced and each may be of kappa or lambda light chain type.[1,2] Multiple myeloma presents in the disseminated form, affecting several bones. Mostly, it is characterized by the multicentric proliferation of plasma cells in the bone marrow. The peak incidence of MM is age around 70 years, and is a rare entity in young patients who are younger than 30 years with less than 2% cases occurring in patients under the age of 40 years.[3] Bone pain involving the back, chest, or extremities are the most common presenting symptoms in MM occurring in 68 to 80 percent of patients. Headache is a rare symptom in MM.

Here, we are presenting a case report of 55 years old female patient with diagnosis of MM who presented with a common presentation low back pain but with a rare presentation as headache. Increased level of β_2 microglobulin, serum creatinine, and plasmablastic type myeloma cells suggest poor prognosis of myeloma patients.

The Case

A 55-year-old house wife presented to outpatient department of Orthopedics of North Bengal medical college, Sirajganj, Bangladesh complaining of headache for three months. She had had these headaches off and on for the past three months, which was more severe for last one week. One week prior to the visit the headaches had increased in severity and frequency, and paracetamol and painkillers no longer afforded relief. The headache was described as sharp and throbbing, and occurred at any time of day. It

was situated predominantly over the frontal region of the head, and on occasion it became generalized and radiated to her ears and mastoid processes. It was aggravated by lights and noise and was not relieved by rest. She had anorexia and nausea accompanied by the pain without any history of vomiting. She noticed occasional dizzy spell lasting a few seconds and sometimes suffered from blurred vision which was more for last one week.

She had no fever, sweating, and weight loss but had history of two units of blood transfusion for anaemia without definitive diagnosis. She had history of low back pain but no arthralgia, arthritis, or kidney associated symptoms and signs.

On physical examination, she was obese and in distress from headache. The oral temperature was 37.4^0C, pulse rate 80/minute and regular, blood pressure 120/80 mm of Hg, and respiratory rate 20/minute.

She was moderately anaemic but not icteric. Oral examination revealed a tongue of normal size. There were no petichiae, ecchymoses or, any lumps. Tenderness over back of the whole spine was noted. The breasts, heart, lungs and abdomen were normal.

The head was extremely tender, especially over the vertex, causing her to pull it away from the palpating hand. Extraocular eye movements and the pupils were normal. There was no exophthalmos. On fundoscopy, there were engorged retinal veins suggestive of hypervis-cosity syndrome.

The haematological investigations revealed anemia (Hb -7 gm/dl), ESR-130 mm in 1^{st} hour, and total WBC count was 13,000/cmm with Neutrophils-85%. Serum creatinine was normal, alkaline phosphatase was increased to 972U/L.

On peripheral smear examination rouleax formation of RBCs were revealed. Serum β_2 microglobulin level was increased and serum albumin was slightly low. Serum protein showed decreased albumin levels, globulin high with A:G reversal (A:G: 1:1.6). On urine routine and microscopic examination Bence Jones proteinuria was absent.

Immunoelectrophoresis and immunofixation studies could not be carried out due to unavailability of these tests and low economic condition of patient. X-ray of the skull (Figure 1), X-ray dorso-lumbar spine (Figure 2) and X-ray pelvis (Figure 3) showed multiple lytic lesions.

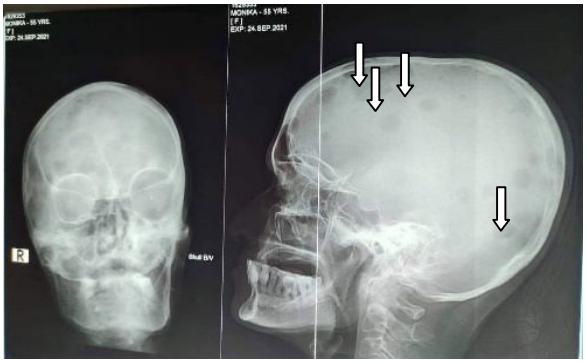

Figure 1: X-ray skull showing lytic lesions

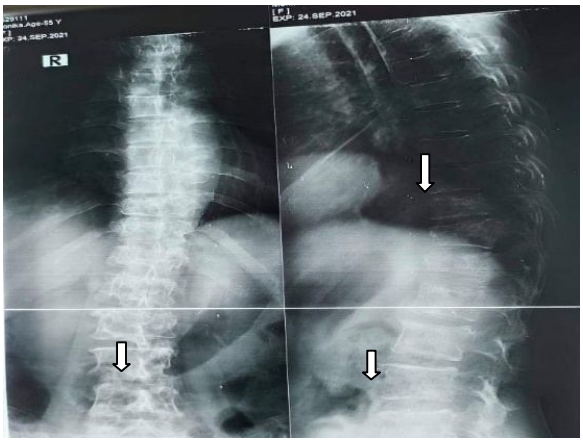

Figure 2: Lytic lesion eroding multiple vertebrae with fractures

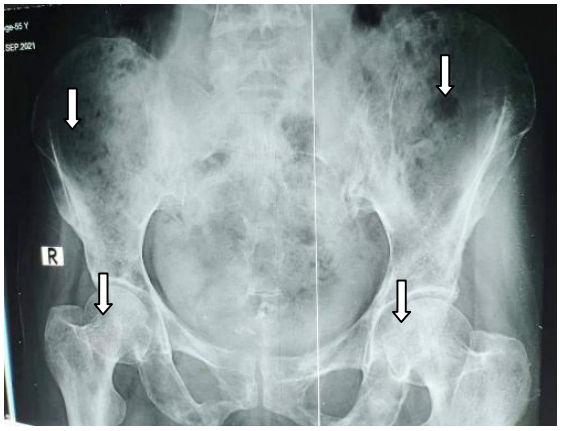

Figure 3: Lytic lesion eroding pubic ramus and femur

Serum biochemical examination revealed: serum creatinine 1.2 mg/dL, urea 25 mg/dL and uric acid 7.40 mg/dL. On serum protein electrophoresis, presence of Beta-2 peak was noted with total protein 3.6 g/dL. There was hyper cellular

marrow and the plasma cells were frequent in bone marrow examination.

On the basis of above findings, this case was diagnosed as Multiple Myeloma. Unfortunately this patient was expired before starting of treatment.

DISCUSSION

Multiple myeloma (MM) is a malignant neoplasm of plasma cells leading to monoclonal proliferation of paraprotein. Usually, few plasma cells are found in the bone marrow and they constitute an important part of body's immune system, working together with other several types of cells to fight infections.[1,2] Most cases of MM patients are often asymptomatic. However, in advanced case, as the disease progresses, patients present with features of infections like fever, fatigue, weight loss, paresthesias, hyper-calcaemia, hyperviscosity, renal failure, bone pain or pathological fractures, cutaneous lesions, etc.

Our patient presented with long standing headache and back pain because of hyperviscosity and multiple lytic lesions in the skull, spine and pelvic bone. Many authors[2, 3, 4] reported no headache in their case of MM but one author[5] reported headache in two patients.

Other common presentations includes renal failure due to dehydration, painkillers, light chain deposition, anemia, fatigue, and hyperca-lcemia.[3,4,5] The median age at diagnosis was 60 to 70 years and the disease is more common in people of African Caribbean origin.[6] Unlike myeloma, Waldenström macroglobulinaemia is a low-grade lymphoplasmacytic lymphoma associated with an Ig M paraprotein, causing clinical features of hyperviscosity syndrome. It is a rare tumor occurring in old age and more commonly affects males. Patients classically present with features of hyperviscosity, such as nasal bleeding, bruising, delirium and visual disturbance. However, presentation is more commonly with anaemia, systemic symptoms, splenomegaly or lymphadenopathy, or may be asymptomatic with an IgM paraprotein detected on routine screening.[6, 7, 8]

Only few patients present with rare features of MM includes headache, hyperviscosity syndrome, amyloidosis, neuropathy.[6] The exact etiology of MM is not known. However, various risk factors like alcohol, obesity have been implicated in its genesis.[9]

Here in our case report, the patient's age was 55 years and we were able to find two unusual features of MM like headache and hyperviscosity syndrome like dizzy spells and visual impairment. Circulating serum immunoglobulins in IgA MM undergo a high degree of polymerization responsible for hyperviscosity and it accounts for 25% cases of hyperviscosity syndrome.[10] Diagnosis of MM requires two of the following criteria to be fulfilled: increased malignant plasma cells in the bone marrow, serum and/or urinary M-protein, skeletal lytic lesions.[8,9]

Haematological analysis of our patient reveals anaemia with marked rouleaux formation because of increased globulins and this is the only reason for high ESR in this patient. Total leukocyte count was increased that indicate infection. Serum β_2 microglobulin level was increased in our case and higher levels are also associated with poor prognosis. On bone marrow examination, hypercellularity was noted due to increased number of plasma cells.

Immunoelectrophoresis and immunofixation studies may demonstrate M band of Ig G/D/A/E. Ig G being the commonest in 60% of the cases. Due to poor socioeconomic condition as well as unavailability these tests cannot be carried out for this patient. Serum viscosity was not analyzed due to unavailability. Measurement of serum viscosity is not required to initiate treatment as because there is no cut-off point for serum immunoglobulins levels that would otherwise indicate the initiation of plasma exchange. The plasma viscosity reduced by 30% to 50% in a single session of plasma exchange and also reduce immunoglobulin level by 60%.[11, 12]

The average survival of patients with MM ranges from two to three years. The survival of the younger patients was considerably longer than patients of other ages with MM.[12] But our patient died before starting of treatment.

CONCLUSION

This case of multiple myeloma represents few rare findings like headache, blurred vision and low back pain. Early diagnosis and management of such cases can increase the length of survival.

Conflict of interest: The authors disclose that they have no conflicts of interest.

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