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Case Report

Extra medullary multiple myeloma of knees: a case report

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ABSTRACT

Multiple myeloma is defined as an incurable, complex, and rare malignant disease of the plasma cells. Multiple myeloma is characterized by uninhibited proliferation of clonal plasma cells. Extramedullary disease is characterized by the presence of clonal plasma cell infiltrate adjacent to soft tissues or distant from the bone marrow in patients with underlying MM. Survival of patients with MM is low. A 65-year-old female reports to the outpatient clinics with pain in the left lower limb for over 2 months. The patient had loss of range of motion. True-cut biopsy revealed multiple myeloma of synovial sarcoma. Extramedullary disease is considered as an uncommon manifestation of multiple myeloma. It is either a newly diagnosed disease or evolves with repeated relapses. There is no clear data on the incidence of extramedullary disease. Since there is a no standard treatment for extramedullary disease, its prognosis remains poor.

Keywords: Extra medullary disease, Myeloma, PET-CT, Plasmacytoma, Prognosis, Survival

INTRODUCTION

Multiple myeloma (MM) is a complex, incurable, and rare malignant disease of the plasma cells. MM is characterized by uninhibited proliferation of clonal plasma cells.[1] MM is associated with diverse complications that often lead to failure and eventually death.[2] Patients with MM often present with renal insufficiency, hypercalcemia, lytic bony lesions, and anemia.[3]

Pathologically, there would be an increase in the number of clonal plasma cells within the bone marrow. The estimated survival term for people with myeloma is between five to seven years, wherein survival is dependent on several prognostic markers.[4] Extramedullary disease (EMD) is defined by the presence of clonal plasma infiltrate adjacent to soft tissues or distant from the bone marrow in patients with underlying MM.[5]

We report an interesting case of extramedullary MM of the knees with soft tissue involvement.

CASE REPORT

A 65-year-old female reports to the outpatient clinic with complaints of pain in the left lower limb in the past 2 months. The patient reported to have swelling, backache, intermittent fever and was unable to bear her bodyweight. The patient on general examination appeared to be stable. No palpable lymph nodes were observed. On local examination, the patient was observed to have a 7x8 cm mass involving the whole left knee. The patient reported having loss of range of motion.

A true-cut biopsy from the soft tissue on the left side of the knee revealed round cell tumours which were indicative of multiple myeloma of synovial sarcoma. As per the immunohistochemistry report, plasmocytoma was observed which was indicative of MM. A bone marrow

aspiration was performed wherein the key histological features favoured MM. Serum protein electrophoresis (SIEP) was carried out with the following results: M band-5.43gm%. As per pathological examination, her B2-microglobulin levels were extremely high, 13165mg/l. ISS staging-III.

The patient was treated with bortezomib and dexamethasone chemotherapy for 24 weeks along with thalidomide 100mg daily. The Patient tolerated chemotherapy very well with no incidence of any cytopenia. The patient showed significant improvement after 8 cycles of chemotherapy. SIEP at eight weeks of chemotherapy was as follows: M band-0.21gm%. The patient responded well to chemotherapy with M band absent after 16 cycles of chemotherapy. The patient developed unexpected weakness after 20 cycles of chemotherapy. Upon pathological investigations, the patient was reported to have pancytopenia. SIEP results were as follows: Two bands observed. The first band observed in junction of beta-2 and gamma region (1.72 gm%) and second band observed in gamma region (0.25 gm%).

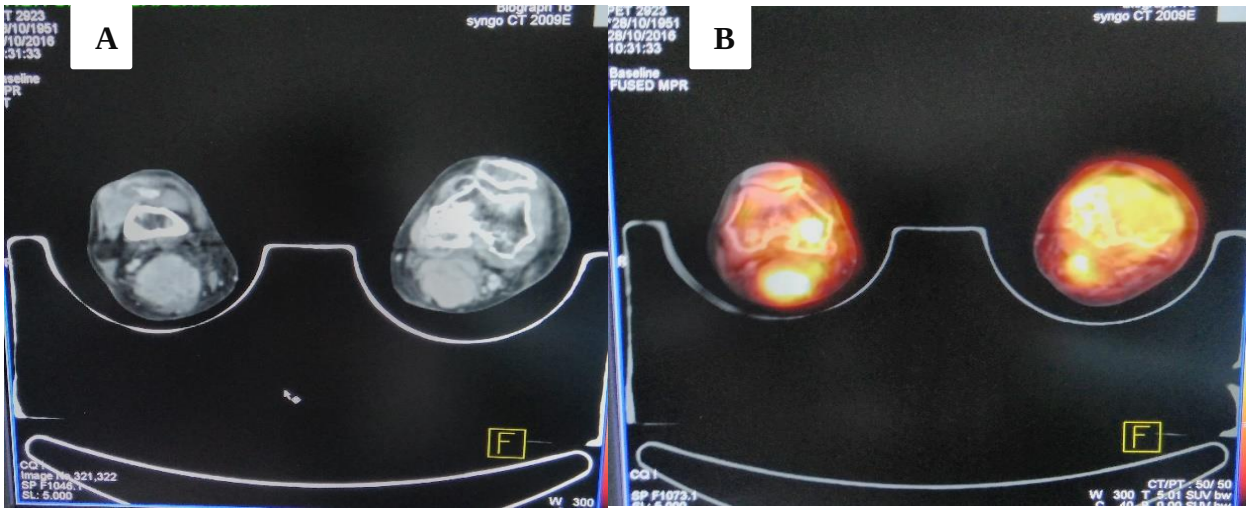

Figure 1: (A) CT Scan- Ill-defined lytic lesion was observed involving upper tibia and tibial plateaue and lower femoral condyles. (B) PET-CT scan- maximum size of lesion measuring 11.7cm x 8.1cm x 7.4cm showing few non-enhancing necrotic foci.

A positron emission tomography (PET) was carried out. As per the radiographs of knee joints, an ill-defined lytic lesion was observed in involving upper tibia and tibial plateau and lower femoral condyles (Figure 1B). Moderate degenerative changes were observed in both knee joints (Figure 1A). Unfortunately, due to severe neutropenia over prolong period and poor general condition patient suffered from pneumonia leading to severe respiratory distress, thrombocytopenia, and grade 4 anaemia after 14 days and eventually died.

DISCUSSION

Multiple myeloma (MM) is associated with the proliferation of malignant plasma cells with a high dependency on the overall bone marrow microenvironment. MM is considered as an exemplar of cancer wherein malignant cells interact with the microenvironment.[6] MM is a known B-cell malignancy that results in fractures and osteolytic lesions. In patients with MM, bone healing is limited due to decreased osteoblastic and increased osteoclastic activity. There is an enhanced tumor progression due to the bone-embedded growth factors due to MM-induced forward-feedback cycle as the bone is resorbed.[7] As per evidence, Extramedullary myeloma (EMM) is preceded by a premalignant disease referred to as monoclonal gammopathy of undetermined significance (MGUS).[8] Nearly 2% of the population aged 50 and above is affected by MGUS, where with a progression to MM at 1% per year.[8]

EMM is considered as a rare manifestation of MM which could be a newly diagnosed disease or evolves with repeated relapses. EMM is characterized by the presence of clonal plasma cells outside the bone marrow in patients with MM.[9] There is limited evidence on the incidence of EM. There is no effective treatment for EMM due to which the prognosis of patients with EM is relatively poor.[10] Based on current evidence, EMD relapses have ranged from 3% to 30%. EMD has been associated with an aggressive form of the disease and short survival rates.[11] The nature of extramedullary involvement is related to disease prognosis. Poor prognosis and outcomes have been observed among

patients with involvement of extra-osseous organs as compared to those with plasmacytomas arising from bones.[11]

CONCLUSION

Despite several developments in the treatment and management of patients with EMM, the prognosis has been poor. There is a need to increase patient participation in clinical trials. A better understanding of the molecular mechanisms associated with the development of EMD would help in developing novel treatment strategies.

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