

Malignant Plasma Cell Disorder in Patients Younger than 20 years

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Abstract

Multiple Myeloma (MM) is a disease of older adults. The occurrence of MM in patients younger than 20 years is rare. In this case-report, we will discuss the story of our 19 year old patient in Syria who presented with isolated bone pain (ostealgia) and compare between old and young patients in both treatment and prognosis of MM. The present findings and the response to conventional therapy in young patients are similar to those in patients of all ages who have MM, however the survival of those patients was considerably longer.

Keywords

Multiple myeloma; Bone-lytic lesions; Younger patients; Malignant

Introduction

Multiple myeloma (MM) is a malignant plasma cell disorder that accounts for approximately 10% of all hematologic cancers [1,2]. The median age at diagnosis is 70 years [3]. Less than 1% of patients are younger than 40 years at diagnosis [4]. Out of 3278, patients reported by Mayo Clinic, 10 patients (0.3%) were younger than 30 years [5]. What makes our case special is that there are only 30 reported cases in literature for MM in patients younger than 20 years [4].

Blade et al. reported a series of 72 patients with MM younger than 40 and 30 years respectively. The frequency of MM in patients younger than 40 and 30 years was 2.2% and 0.3% respectively. We have reviewed 150 Myeloma cases of past ten years who were treated in our center of these only 5 patients (3.3%) were less than 30 years age [4]. In this case-report, we will discuss the story of our 19 year old patient who presented with isolated bone pain (ostealgia) and compare between old and young patients in both treatment and prognosis of MM.

Case Presentation

This is the case of a 19-year-old boy who presented with ostealgia in the knee, right femur, and ribs, which was not relieved by analgesics. He was otherwise healthy, with insignificant medical and family history. Simple X-ray of the skull and pelvis revealed several bone-lytic lesions. The following step in Differential diagnosis was the skeletal scintigraphy which revealed high uptake of technetium Tc99 (Figure 1). The serum protein electrophoresis showed a monoclonal spike on γ globulin. Furthermore, serum IgG was 1745 mg/dl, IgM 119 mg/dl, IgA 225 mg/dl, and β_2 microglobulin 1.3 mg/l. The bone marrow study quantified the presence of plasma cells (Figure 2). Histological examination and the immunohistochemical study of the bone marrow biopsy and opened biopsy of the knee lytic lesion revealed atypical plasma cell infiltration. In addition, the immunophenotyping study detected positive antigens for both (CD138, LCA) and was negative for CD99. Conventional cytogenetics were performed, no evidence of any chromosomal abnormality. There was no evidence of renal failure nor anemia as the lab results suggested (Cr: 0.7, Hgb: 15.7 g/dl, Plt: 159 C/uL, Ca: 9.1 mg/dl, ALP: 65 U/L, ESR: 38 mm). Eventually, the diagnosis of Multiple Myeloma was established on the previous bases. Consequently, induction chemotherapy was initiated with Bortezomib (3 mg weekly for 16 weeks), (Dexamethasone 40 mg weekly for 16 weeks), Lenalidomid (25 mg daily d1-d21) for four 28 days cycles. Then the patient also needed irradiation therapy for his right knee (45 gry). After months, the patient became asymptomatic and bone tumor regression, complete hematological remission confirmed by a normal serum protein electrophoresis and normal bone marrow biopsy, no evidence atypical plasma cell infiltration. The patient is currently programmed for the first autograft.

Discussion

Multiple myeloma is a considerably rare plasma cell proliferation disease, accounting for almost 15% of all hematologic malignancies and 1% of all malignancies. Moreover, diagnosing this cancer in young patients is quite uncommon; approximately 30 cases were reported in the literature of patients younger than 20 years old [4]. In this paper, we discuss the case of a 19-year-old boy in Syria. In most case-reports, presentation in younger

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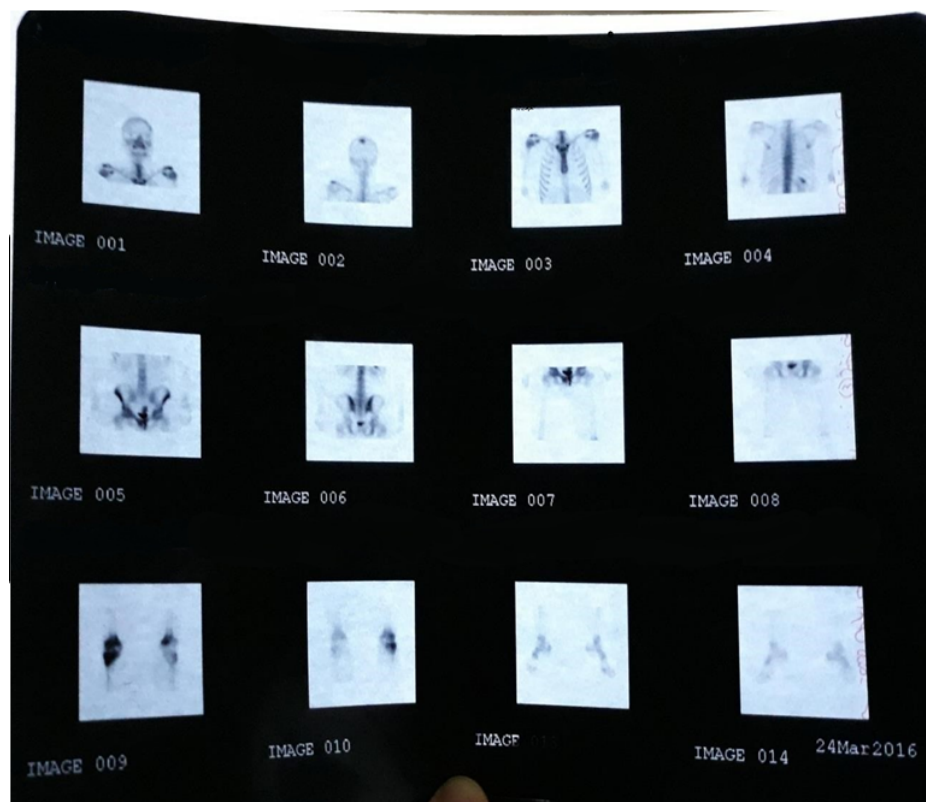


Figure 1: Skeletal scintigraphy revealing high uptake of technetium Tc99

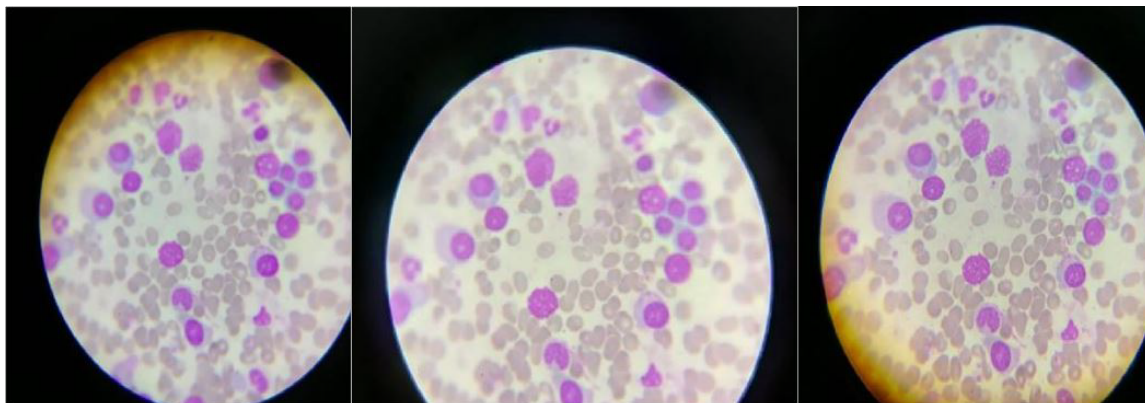


Figure 2: Histopathological examination of bone marrow biopsy (H&E staining)

patients is less severe. Accordingly, our patient suffered from bone-isolated pain, which typically corresponds to bone-lytic lesions. Therefore, the radiologic findings suggested further investigation with Multiple Myeloma on top of the differential diagnosis list. With the bone marrow infiltrated by plasma cells (Figure 2) and the presence of MGUS (Monoclonal gammopathy of undetermined significance), the diagnosis was made based on two major criteria and one minor criterion. However, the patient did not experience kidney involvement, which is a common component of the clinical setting in young patients [4,5]. MM is unusual in the young patients. The clinical course in adolescents and young individuals is generally indolent and the survival is longer however, the response to chemotherapy is variable. Lazarus reported two cases of plasma cell myeloma in young patients, one was a case of MM involving the skull and ribs in a 23 year old woman, the other was a solitary myeloma of the tibia in a 21 years old man. Both the cases were diagnosed non-secretory myelomas [6].

Curose et al. reported the first use of Bortezomib according to

their 8-year-old child case report in 2014 [5]. Unfortunately, although many trials are being conducted regarding the use of Bortezomib to treat pediatric neoplasms, no certain information is available in the literature about using this medication in children [7]. Considering all this, we tried to follow IMWG treatment guidelines within the available resources in Syria currently (2017-2018). Hence, the patient received a combination of Bortezomib, bethamethazone, and lenalidomide since several studies suggested the superiority of three-drug bortezomib-based regimen for induction therapy (IMWG) [8]. In addition, he received irradiation therapy for knee lesions. Few months later, all clinical symptoms improved and the patient moved to an asymptomatic phase. Transplantation options were discussed since we have complete remission. In the study from Mayo clinic, the median duration of survival of the patients was 87 months the survival of the younger patients was considerably longer than that of patients of all ages with MM. Thus, the occurrence of myeloma in the younger individual does not appear to impart a worse prognosis or survival [9,10]. Last but not least, Geetha et al. did a small-lit review

on the clinical features of MM in young patients until 1999, do you want to proceed and conduct a new review on all patients until our time? This can make it a case report and a literature review.

Conclusion

The case findings and the response to conventional therapy in young patients are similar to those in patients of all ages who have MM, however the survival of those patients was considerably longer.

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