



Case Report of Multiple Myeloma Presenting with Unique Skin Manifestation with Literature Review

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Abstract

Multiple myeloma (MM), although a rare disease, is the second most common hematologic malignancy. MM is associated with significant morbidity due to its end-organ destruction. It is a disease of the older population and advancements in the diagnosis, monitoring, and treatment of MM are of the utmost importance as the general population lives longer due to other improvements in health care. Herein, we present an interesting case of MM presenting with initially skin manifestation which unfortunately is associated with a poor prognosis due to a relationship indicating higher tumor burden. Despite being on multiple treatment, our patient continued to progress with her disease.

Introduction

Multiple myeloma (MM) accounts for approximately 2% of all malignancies and 13% of hematological malignancies worldwide [1]. MM is the second most common hematologic malignancy [1] and it is a neoplasm of clonal plasma cells which originate from the B-cell lineage and develop after lineage commitment in the bone marrow of progenitor cells [9]. Cutaneous involvement in patients with multiple myeloma is rare and it usually represents of a poor prognosis. Cutaneous involvement indicates an increased tumor burden. The most common cause of cutaneous involvement is due to direct extension from underlying bone lesions of MM or solitary plasmacytoma of bone [2]. It is very rare to have primary cutaneous plasmacytoma [3].

Case Presentation

We present the case of a friendly Caucasian American 70-year-old female with no significant PMH who initially presented to Ruby Memorial Emergency Department in November 2015 with persistent back and left side pain. At that time, she began to notice weakness and some numbness in lower extremities and along with the increasing low back pain decided to seek medical attention and presented to the hospital. On admission, CT thoracic spine (11/16/15) revealed a T5 wedge compression deformity.

CT lumbar spine revealed degenerative changes with diffuse osteoporosis. MRI thoracic spine (11/17/15) revealed a T5 burst fracture with retropulsion, focal kyphotic deformity, severe central canal stenosis, possible pathologic fracture deformity, And possible cord contusion at T5. CBC on admission revealed WBC 5.1 with normal differential, Hgb 11.5 with MCV 105.3 and platelet count 170,000. BMP normal, creatinine 0.75, corrected calcium 10.2. Subsequently, she underwent a posterior spinal fusion T3-T8, left costotransversectomy with left pedulectomy and subtotal corpectomy (excision of tumor) via transpedicular approach with placement of anterior cage, T5 laminectomy, and T6-7 partial laminectomy. Pathology revealed a tumor composed of sheets of atypical plasmacytoid cells positive with immunostain for CD138 and negative for pancytokeratin, CD20, and CD3. Insufficient tissue remained for kappa and lambda ISH. The finding was consistent with plasma cell neoplasm. SPEP revealed an M spike seen in the gamma, consistent with monoclonal gammopathy. Random UPEP/IFE revealed a monoclonal lambda protein without detectable associated IgG, IgA or IgM heavy chain. In December, SPEP/IFE revealed an IgG Lambda monoclonal protein (5.95 g/dL). Free light chain ratio 0.013 (Kappa 0.80 mg/dL, Lambda 60.50 mg/dL). Quantitative IgG 5950 g/dL. Hgb 9.4 with MCV 98.5. Creatinine 0.62.

Corrected serum calcium 11.2. Albumin 2.0. Beta 2 microglobulin 4.25. Skeletal survey reveals diffuse osteolytic process with multiple calvarial, vertebral body and rib lesions identified. T3-T8 posterior spinal fusion for pathological T5 compression fracture noted.

She had a bone marrow biopsy/aspirate which revealed 47% lambda restricted plasma cells, with marked atypia including a plasmablastic morphology. Preliminary FISH results indicate multiple trisomies, 1q21 gain, del (13q14) and a non-standard IGH rearrangement-t(8;14). Hyperdiploidy detected. She was diagnosed with IgG Lambda Multiple Myeloma. ISS Stage II, DS IIIA. Soon thereafter, she was started on a bortezomib-based triplet regimen - bortezomib, cyclophosphamide, dexamethasone (CyBorD). She completed 4 cycles. In April 2016, SPEP/IFE revealed an IgG Lambda monoclonal protein - not quantified. Quantitative immunoglobulins: IgG 1013 mg/dL. Free light chain assay revealed a ratio of 1.169 (kappa 2.98 mg/dL, Lambda 2.55 mg/dL). CBC normal. Hgb 13.2. Serum creatinine 0.59. Serum calcium 9.7 (corrected). Serum albumin 3.0 g/dL. Beta 2 microglobulin 2.65 mcg/mL. Skeletal survey demonstrated new evidence of L5 pathological compression fracture. A repeat bone marrow biopsy revealed a variably cellular marrow (10-50%). Maturing trilineage hematopoiesis noted. No atypical plasma cell infiltrate (<5% by CD138 ICH). Cytogenetics normal. Unable to run FISH. At the end of the month, her regimen switched to Lenalidomide/Dexamethasone and she completed 4 cycles. In August, she began maintenance therapy with Lenalidomide at 10 mg daily. She then presented with multiple skin papule skin lesions (Figure 1) mostly on her gluteal region. In January 2017, she had a left gluteal skin biopsy consistent with plasma cell neoplasm with high grade features (plasmablastic differentiation) In February 2017, SPEP/IFE revealed a IgG Kappa monoclonal protein (not quantified). IgG level 2303 mg/dL. Free light chain assay reveals a ratio of 1.31 (Kappa 12.63 mg/dL, Lambda 9.62 mg/dL). She completed radiation therapy to left gluteal skin - received 30 Gy/15 fractions. SPEP/IFE revealed a IgG Kappa monoclonal protein (not quantified). IgG level 2357 mg/dL. Free light chain assay reveals a ratio of 1.47 (Kappa 12.20 mg/dL, Lambda 8.32 mg/dL). In March 2017, she was started on Pomalidomide/Dex. Unfortunately she was found to have progression of her disease noted. She was then started on Daratumumab which she could not tolerate and was then started on Carfilzomib/Dex. She has received multiple radiation therapy to her left gluteal region, mid back and left upper lip with significant resolution in all areas. In February 2017, she was admitted to Ruby with worsening pain and lesions located in her lower extremities and edema (Figure 1). Peripheral duplex was obtained and blood cultures to rule out a venous clot and infection respectively. A biopsy was obtained (Figure 2) which showed cutaneous involvement of her multiple myeloma. Radiation Oncology was consulted, and she was started on palliative radiation for 5days which initially improved her pain

and swelling. Unfortunately, her lesions continue to arise with intermittent, short lived responses to systemic therapy. Her lesions now too widespread for continued radiation therapy. She noticed lesion in her scalp. She continues to follow up in clinic where she was started on single agent Doxil 40 mg/m² every 4 weeks. Sadly, she continues have progressive cutaneous disease around the flank, abdomen, and bilateral thighs.



Figure 1: Skin Manifestation.

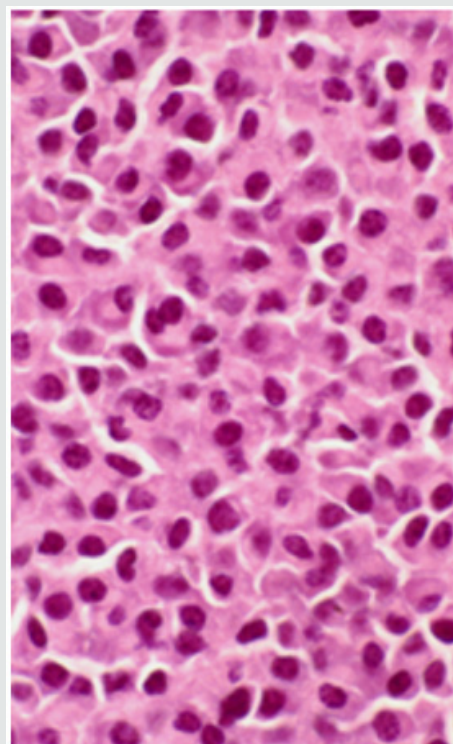


Figure 2: Nodular Pattern.

Discussion

We present an interesting case of cutaneous involvement of multiple myeloma where although the patient has been on multiple therapies continues to progress in her disease. Her case is interesting from other presentations in that they don't usually do not describe the course of regimen used in treatment. The standard of care of cutaneous involvement revolves controlling the origin of the disease. One specific treatment that we tried was localized radiation. There is not much data or past literature discussing the use of radiation or its efficacy. Unfortunately for our case, the treatment only helped briefly. Most common involvement for MM is soft tissue involvement of the upper airway and oral cavity. They usually consist of firm, erythematous, nontender nodules involving the neck, ears, shoulders, axillae, chest, abdomen, and dorsum of the hands [11]. The first reported case of skin involvement in a person with MM was presented by Bruno Block in 1910. He described a patient who had small reddish macules that evolved into brown reddish papules and nodules with scale crusts. Histologically these lesions showed epidermal necrosis. He eventually had disease in pleura, stomach, and heart and passed away two years later [3]. A review of literature reveals that there are over 100 described cases. The age ranges from 36 to 81 with a median of 60 years old. Numeric date was available for 87 cases and 63 of them were male and 24 were female [4]. Cutaneous involvement of MM may appear in area of the skin, but it has been reported most commonly on the

trunk and abdomen. Skin lesions is commonly described as papules or nodules that measure 1-5 cm in diameter with firm consistency, smooth surface, and a red or violaceous color [5]. Some authors reported that cutaneous involvement of MM only occurs when the tumor burden is over 2-3 kg [6]. Cutaneous involvement in patient with MM and extramedullary plasmacytoma generally appears late during the disease. On average, death occurred within 12 months after the diagnosis. Autopsy of these patients reveal extensive plasmacytic infiltration of multiple organs [7].

A review of the cases of MM involving the skin revealed that 40 cases were IgG, 21 cases were IgA, and 9 cases were IgD. The risk of cutaneous involvement by MM is not associated with a particular class of myeloma immunoglobins. Histopathologically, the lesions of MM involving the skin show 2 patterns: nodular and diffuse interstitial [6]. (Figure 2, 3). The worldwide incidence of myeloma is 86,000 cases annually. Mortality rate in MM is high with a median survival of 50-55 months and 63,000 deaths being reported worldwide each year [8]. Significant advances have been made in understanding multiple myeloma (MM) and its precursor diseases. These advances include the gain in knowledge in the underlying pathophysiology, Food and Drug Administration (FDA) approvals of novel therapies with meaningful efficacy and the science in underlying disparities in patients with MM [9].

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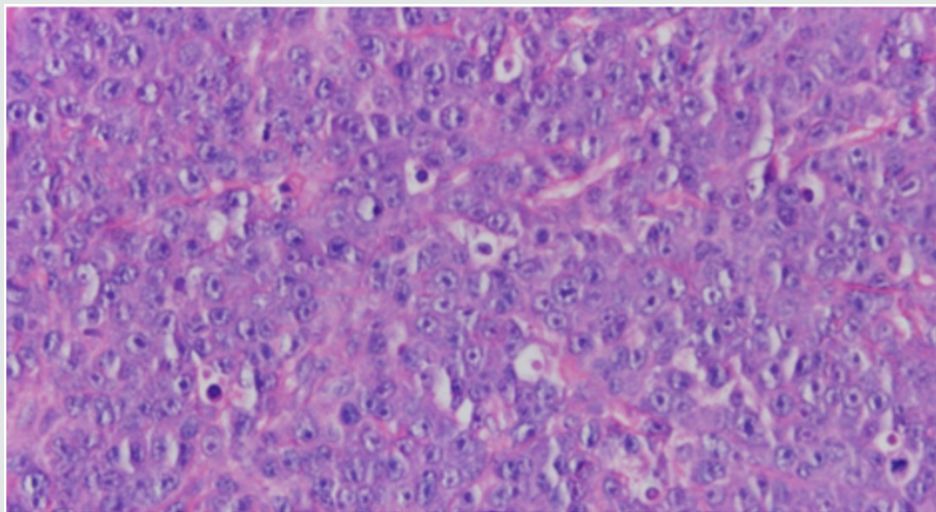


Figure 3: Diffuse Pattern.

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