Open Access Library Journal
2025, Volume 12, e12574
ISSN Online: 2333-9721
ISSN Print: 2333-9705

Multiple Myeloma in Young African Woman with Systemic Lupus Erythematosus and Sjögren Syndrome: A Case Report

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How to cite this paper: Sabounji, M.M. and Ndiaye, C. (2025) Multiple Myeloma in Young African Woman with Systemic Lupus Erythematosus and Sjögren Syndrome: A Case Report. *Open Access Library Journal*, **12**: e12574.
https://doi.org/10.4236/oalib.1112574

Received: November 6, 2024
Accepted: January 6, 2025
Published: January 9, 2025

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Abstract

Background: Systemic lupus erythematosus (SLE) and Sjögren Syndrome (SS) are chronic autoimmune diseases with a spectrum of clinical and serological presentations. Multiple myeloma (MM) has been demonstrated in the course of these diseases. However, the occurrence of MM in young patients under 30 years with SLE and SS has never been described. **Case presentation:** We describe the first documented case of a 27-year-old Senegalese woman who presented MM after two years with SLE and SS symptoms. The diagnosis of these conditions was made simultaneously at presentation. The diagnosis of SLE associated with SS was made based on the presence of joint involvement, acute cutaneous lupus, non-scarring alopecia, positive antinuclear antibody, positive anti-Smith antibody for SLE and positivity of anti-SSA (Ro) antibodies, positive Schirmer tear test for SS. On the other hand, the diagnosis of MM was made based on inflammatory bone pain, peak monoclonal hyper gammopathy, bone lytic lesions on skull radiography, and bone marrow aspiration revealing plasma cells at 7% with some dystrophic plasma cells. Due to a lack of resources, immunofixation electrophoresis was not performed. **Conclusion:** Systemic lupus erythematosus and Sjögren's Syndrome increase the risk of multiple myeloma. The clinicians should consider the possibility of multiple myeloma in young patients with these autoimmune diseases.

Subject Areas

Rheumatology

Keywords

Multiple Myeloma, Systemic Lupus Erythematosus, Sjögren Syndrome, Young

1. Background

Systemic lupus erythematosus (SLE) and Sjögren Syndrome (SS) are chronic autoimmune diseases with a spectrum of clinical and serological presentations. These two conditions can be accompanied by a monoclonal gammopathy of undetermined significance (MGUS) and rarely can be associated with the development of multiple myeloma (MM) [1]. On the other hand, multiple myeloma is a B-cell malignancy characterized by excess expansion of malignant plasma cells in bone marrow [2]. Multiple myeloma is extremely rare in patients less than 30 years of age [3]. The occurrence of MM in young patients under 30 years with SLE and SS had never been described. Herein, we report the first documented case of multiple myeloma in a young African woman with SLE and SS.

2. Case Presentation

A 27-year-old Senegalese female presented with a two-year history of polyarthralgia and Sicca symptoms. She reported the aggravation of these symptoms during the last 3 months by apparition of inflammatory bone pain including skull and whole spine, fatigue and weight loss. She took non-steroidal anti-inflammatory drugs and analgesics in primary health facilities without clinical improvement.

On admission, vital signs including body temperature were normal. Physical examination revealed peripheral polyarthritis (without deformities or ankyloses) and myalgia with muscle weakness (muscle testing: grade 4). She also presented butterfly rash, moderate alopecia of the scalp and Sicca symptoms (dry eye and dry mouth). Neurologic examination demonstrated no signs of compression of the spinal cord or nerve roots. She had no other symptoms including Raynaud's phenomenon, Gottron's papules, dyspnea, oral ulcers, parotidomegaly, splenomegaly, hepatomegaly, lymphadenopathy, or night sweats. She also had no digestive symptoms (nausea, vomiting, dysphagia, dyspepsia, epigastric pain, gastric bleeding).

Laboratory findings showed normochromic normocytic anemia at 10.4 g/dl without other abnormalities on the blood count test, elevated serum C-reactive protein at 24 mg/l (normal < 6), raised erythrocyte sedimentation rate at 35 mm/hour and elevated Creatine Kinase at 756 IU/L (normal < 170). Antinuclear antibody was highly positive at 1/1280 (positive > 1/80) with a speckled pattern. Anti-Smith, anti-SSA (Ro), anti-SSB (La) and anti-U1 RNP antibodies were positive. However, anti-CCP, Rheumatoid Factor, anti-DNA, anti-centromere and anti-Scl70 were negative. Otherwise, Myositis-Specific Antibodies were negative. Serology of HIV, Hepatitis B, and Hepatitis C were negative.

Other laboratory results including creatinine, glomerular filtration rates, calcaemia, 24-hour urinary protein and liver function tests were within the normal limits.

The diagnosis of SLE was made according to the 2019 ACR-EULAR classification criteria for SLE: presence of joint involvement (6 points), acute cutaneous lupus (6 points), non-scarring alopecia (2 points) and anti-Smith antibody (6

points).

We also diagnosed Sjögren Syndrome based on xerophthalmia with a positive Schirmer tear test ≤ 5 mm/5min and positivity of anti-SSA (Ro) antibodies. Thus, our patient fulfilled the 2016 ACR-EULAR classification criteria for SS with a total score of 4 points: positivity of anti-SSA (Ro) antibodies (3 points) and positive Schirmer test ≤ 5 mm/5min (1 point). The patient also presents xerostomia; however, a labial salivary gland biopsy with histology was not performed due to a lack of resources.

On the other hand, the patient presented a three-month history of inflammatory bone pain including skull and spine with weight loss. Radiography of the skull showed diffuse demineralization with a lytic lesion (Figure 1). Serum protein electrophoresis showed peak monoclonal hypergammopathy at 4.4 g/dl. Bone marrow aspiration revealed plasma cells at 7% (Normal range: 1% - 2%) with some dystrophic plasma cells. Serum and urine immunofixation electrophoresis were not performed due to insufficient resources.

A diagnosis of multiple myeloma was made based on inflammatory bone pain, peak monoclonal hypergammopathy, plasma cells at 7% and lytic bone lesions on skull radiography.

Otherwise, there was no evidence of lymphoma.

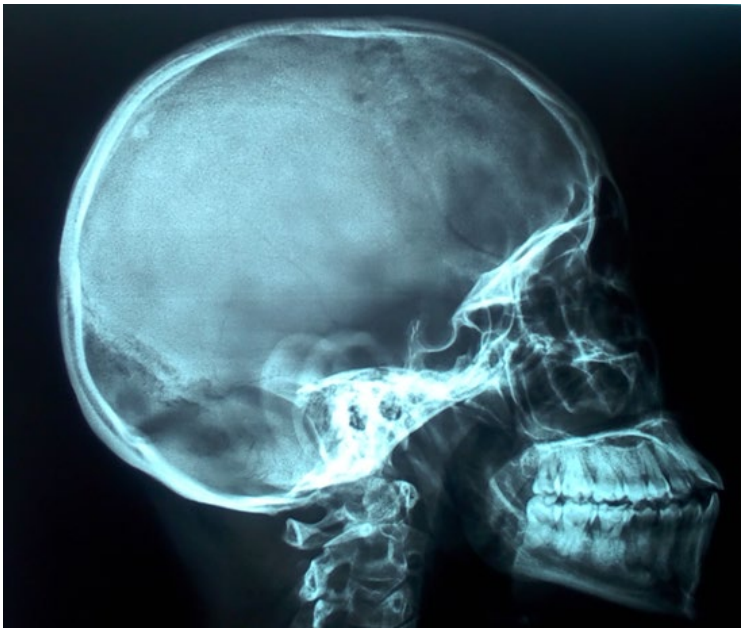

Figure 1. Radiography of skull showing bone lytic lesions.

We started therapy for SLE and SS including Hydroxychloroquine (400 mg daily), Methotrexate (15 mg per week), Prednisone (15 mg daily), Triamcinolone acetonide 80 mg intra-articular injection on knees. On the other hand, she received Zoledronic acid 4 mg intravenously for lytic bone lesions (multiple myeloma-related bone disease). Then, the patient was referred to the hematology department to start chemotherapy for MM.

3. Discussion

We describe an unusual case of multiple myeloma in a young African woman with systemic lupus and Sjögren Syndrome. To our knowledge, this is the first documented case in literature.

A meta-analysis showed a significantly increased risk of multiple myeloma in SLE patients compared with the general population [4]. Several theories have been suggested to explain the association of MM and SLE: B cells' hyperactivity in SLE which favors the escape of abnormal B cell clones from the normal regulatory mechanisms; defective immunologic surveillance of malignant cells in SLE; increased resistance to cell apoptosis may further promote the malignant transformation of tumors; mutations in the phosphatase and tensin homolog (PTEN) gene and Bcl-2 overexpression may be the basis for resistance to cell apoptosis [5] [6].

A total of 15 cases of SLE associated with MM have been reported cumulatively from 2000 to 2022 [6]. The average age at the time of SLE diagnosis was 40.07 years (age range, 22 - 76 years). The average age at the time of MM diagnosis was 50.80 years (age range, 28 - 76 years). Except for one patient in whom MM and SLE were diagnosed simultaneously, multiple myeloma occurred after SLE in all other cases.

Regarding Sjögren Syndrome, a meta-analysis showed a significantly increased risk of multiple myeloma in Sjögren's Syndrome patients [7]. Moreover, mofors *et al.* observed increased risk of multiple myeloma in primary sjögren's Syndrome is limited to individuals with Ro/SSA and La/SSB autoantibodies [8]. Our patient had positivity of these autoantibodies. In SS, the B cell stimulation is not restricted to B cells and continues until the final differentiation; plasmablasts and plasma cells are increased in the blood and salivary glands, respectively [9]. This hyperactivity of plasmablasts is probably predisposing to multiple myeloma.

A total of 9 cases (8 females and 1 male) of SS associated with MM have been reported in the literature [10]-[18]. The mean age at the time of SS symptoms/diagnosis was 55.7 years (age range, 41 - 72 years). The mean age at the time of MM diagnosis was 61 years (age range, 41 - 72 years) [10]-[18].

Although the diagnosis of multiple myeloma requires the presence of a plasma cell count above 10%, this condition is not necessary if there is end-organ damage (hypercalcemia, renal failure, anemia and bone lesions). According to a Mayo Clinic study [19], 4% of patients with multiple myeloma have bone marrow plasma cells less than 10%. Our patient presented bone marrow plasma cells at 7% with lytic bone lesions.

The pathogenesis of lytic bone lesions in MM constitutes a multifaceted entity that includes several intracellular and intercellular signaling pathways [20]. The intercellular interactions between bone marrow stromal cells (BMSCs) and MM cells along with the involvement of immune cells, such as Th17 cells, induce cytokine release (IL-1b, IL-3, IL-6, IL-11, IL-17) and secretion of osteoclastogenic factors (OAFs) such as TNF-α, CCL-3, SDF-1α, and Annexin II in the bone marrow

microenvironment. These cytokines promote increased osteoclast activity [20]. OAFs were initially identified in conditioned media from myeloma cell lines and found to stimulate bone resorption in bone organ culture systems [21]. Interestingly, several of these OAFs also suppress osteoblast formation (inhibit osteoblastogenesis) and/or support myeloma cells directly, indicating that they play multiple roles in MM bone disease [22]. Adhesion molecules such as VCAM-1 on BMSCs and VLA-4 on MM cells mediate cell-to-cell contact. Notch, expressed by MM cells, binds to Jagged, expressed by neighboring MM cells and BMSCs, and activates intracellular cascades favoring RANKL production. MM cells also enhance the apoptosis of osteocytes that also release RANKL. RANKL binds directly to RANK on osteoclast precursors and promotes osteoclastogenesis [20].

The occurrence of multiple myeloma in our patient at such a young age was mainly linked to two aspects: firstly, the concomitant presence of systemic lupus and Sjögren Syndrome increases the risk of multiple myeloma development. Secondly, the delayed diagnosis of systemic lupus and Sjögren Syndrome. Our patient spent two years with these two autoimmune diseases in primary care facilities without specific treatment for these conditions.

In fact, early diagnosis and adequate treatment of SLE and SS would have prevented the development of tumor complications such as myeloma.

4. Conclusion

We described the first case reported in the literature of multiple myeloma in a young African woman with Systemic lupus and Sjögren Syndrome. The clinician should consider multiple myeloma in young patients with these autoimmune diseases.

Conflicts of Interest

The authors declare no conflicts of interest.

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