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Case Report

From sicca to malignant evolution: a swift turn to multiple myeloma in primary Sjögren's syndrome

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ABSTRACT

Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder characterized by exocrine gland involvement and systemic B cell hyperactivity. Although hematologic complications are recognized, transformation to multiple myeloma (MM) remains exceptionally uncommon. We report a 50-year-old woman who presented with constitutional symptoms, sicca manifestations and recurrent vasculitic skin lesions. Evaluation established a diagnosis of primary Sjögren's syndrome with associated monoclonal gammopathy of undetermined significance (MGUS). Despite initial management and follow up, she subsequently developed worsening systemic symptoms accompanied by progressive extra glandular manifestations, prompting reassessment. Serial investigations confirmed progression to multiple myeloma within a relatively short interval. The patient was managed collaboratively by rheumatology and hematology departments and showed favorable clinical and laboratory response to therapy. This case highlights a rare malignant evolution in pSS and underscores the importance of maintaining a high index of suspicion in patients with persistent systemic symptoms, paraproteinemia, vasculitic manifestations or unexplained clinical deterioration. Careful longitudinal monitoring may facilitate earlier recognition of hematologic transformation and enable timely therapeutic intervention, potentially improving patient outcomes.

Keywords: Primary Sjögren's syndrome, Multiple myeloma, Vasculitic skin lesions

INTRODUCTION

Primary Sjögren's syndrome (pSS) is a chronic autoimmune disorder having diverse clinical manifestations, ranging from sicca symptoms, and fatigue to systemic complications due to chronic B-cell activation, hypergammaglobulinemia, and an increased risk of lymphoproliferative disorders, particularly mucosa-associated lymphoid tissue (MALT) lymphoma.[1] MGUS and its progression to multiple myeloma (MM) in pSS is rare and risk factors for transformation include cryoglobulinemia, hypergammaglobulinemia, lympha-denopathy, auto antibody positivity (anti-Ro/SSA, RF), older age and low complement levels.[2] Although MGUS is frequently benign, it can progress to MM or other lymphoid malignancies in about 1% of cases per year.[3] This case illustrates a unique and rapid transformation of MGUS to MM within a year in a pSS patient, adding to the limited literature on such progression in the context of Sjögren's syndrome.

CASE REPORT

A 50-year-old postmenopausal woman with constitutional symptoms over five years and oral sicca symptoms presented with a sudden-onset skin rash characterized by tender erythematous lesions over her arms and legs following an episode of gastroenteritis which later progressed to involve other areas of the body, leading to extensive desquamation and non-healing, painful ulcers over the dorsum of the foot with the evidence of vasculitis on histopathology. Autoantibody testing revealed strongly

positive ANA [IF] + 4 intensity, speckled pattern in 1:100 dilution with SSA positivity [+3] and borderline SSB positivity. Further evaluation showed pancytopenia, elevated globulins with A/G reversal, and elevated ESR. A suspicion of pSS with possible cryoglobulinemia or paraproteinemia was considered. Schirmer's test suggests severe dryness of both eyes and minor salivary gland biopsy suggestive of Sjogren's syndrome with a focus score >1. Protein electrophoresis demonstrated an M-spike of 1 g/dl and elevated serum-free light chains kappa light chains of 441 mg/l (normal range 3.3-19.4 mg/l), lambda light chains of 134 mg/l (normal range 5.7-26.3 mg/l), and an elevated Kappa/Lambda ratio of 3.29 (normal range 0.26-1.65). Cryoglobulin testing by cryo kit (qualitative method) was negative. Bone marrow biopsy showed 8% plasma cells with CD138 positivity.

A diagnosis of pSS with MGUS was made and treated accordingly with glucocorticoids after initial hematology consultation. In the next year, she presented with worsening constitutional symptoms, sicca symptoms, and paresthesias of both lower limbs, along with recurrent oral ulcers and painful erythematous maculopapular skin lesions over lower limbs that progressed to ulceration over the feet and lower third of the legs with necrotic patches (Figure 1).

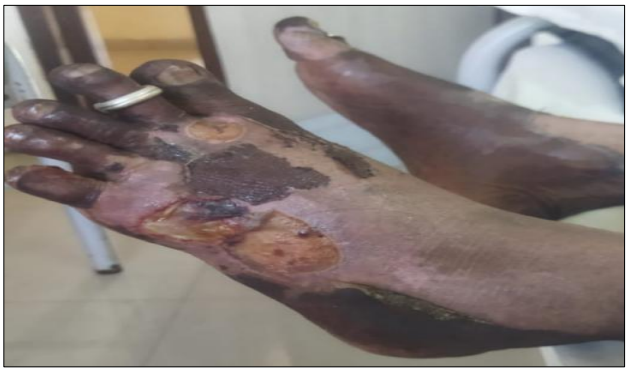

Figure 1: Desquamation and ulcers.

On examination, she had digital tip infarcts on the toes and fingers (Figure 2), oral thrush, glossitis, impaired fine touch, and vibration sensation over both lower limbs below the knee with decreased ankle reflexes. Further laboratory tests showed persistently elevated globulins with A/G reversal. Complement studies revealed normal C3 levels but significantly reduced C4 levels 6 mg/dl (10-40 mg/dl).

Nerve conduction studies showed symmetric sensory axonopathy. Serum electrophoresis demonstrated increased levels of kappa and lambda light chains, kappa - 904 mg/l (normal range 3.3-19.4 mg/l), lambda - 800 mg/l (normal range 5.7-26.3 mg/l) compared to previous values, raising suspicion of transformation to multiple myeloma and repeat bone marrow biopsy showed increased plasma cells to 30% with CD 138 positivity (Figures 3 and 4).

The skeletal survey showed no lytic lesions. Hence, a diagnosis of pSS with MGUS showing rapid transformation to multiple myeloma was made.

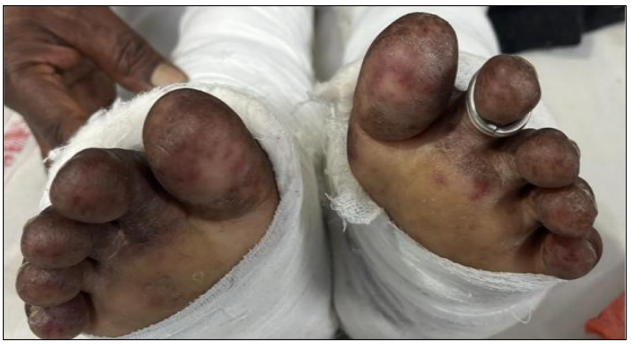

Figure 2: Digital tip infarcts.

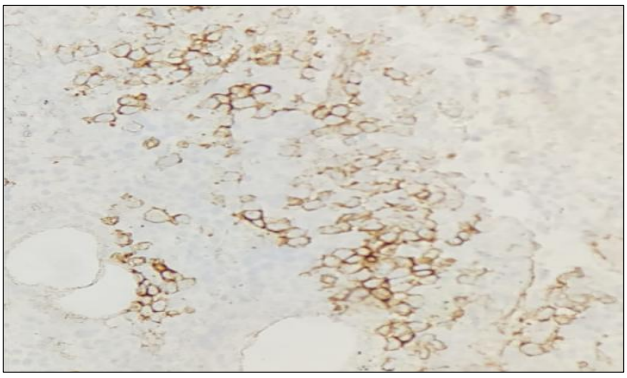

Figure 3: CD138 stained plasma cells.

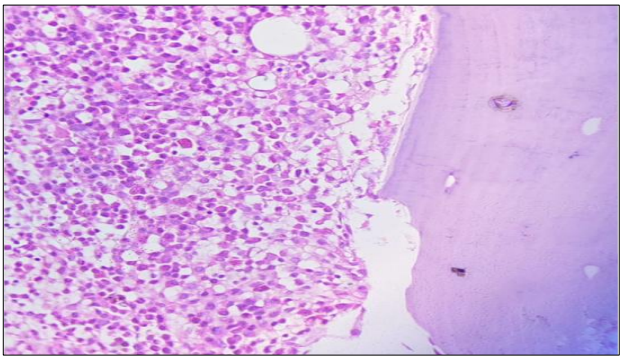

Figure 4: Increased marrow cellularity.

The patient was initiated on weekly intravenous cyclophosphamide, bortezomib and dexamethasone 0.5 mg/kg in consultation with hematology and the patient was followed up over the next 6 months after initiation of bortezomib-based chemotherapy. She showed significant clinical improvement with resolution of constitutional symptoms, healing of cutaneous ulcers, and no new vasculitic lesions. Neuropathic symptoms stabilized with partial improvement in sensory deficits. Serial laboratory monitoring demonstrated a reduction in serum free light chains and improvement in globulin levels. No new end organ damage or lytic lesions were detected on follow up

evaluation. The patient continues to remain on regular follow up under rheumatology and hematology departments with ongoing therapy and monitoring for disease response and complications.

DISCUSSION

Patients with pSS are known to be at increased risk for B cell malignancies, particularly MALT lymphoma and other lymphoproliferative disorders such as Waldenström's macroglobulinemia and monoclonal gammopathy of undetermined significance (MGUS), but the progression of MGUS to multiple myeloma in pSS is rare.[1,4] pSS is characterized by B-cell hyperactivity and lymphocytic infiltration of exocrine glands, with approximately 5% of patients developing B-cell lymphomas.[5] A state of chronic immune stimulation is believed to contribute to lymphomagenesis, and patients with SS frequently exhibit benign monoclonal gammopathy in serum or urine, although progression to multiple myeloma remains rare.[6] Chronic inflammation has been proposed as a potential driver in the evolution of plasma cell dyscrasias, suggesting that the underlying immunological dysregulation in SS may also contribute to the development of monoclonal gammopathy.[7]

The patient initially presented with hypergamma-globulinemia, a typical feature of Sjögren's, Mspike and bone marrow 8% plasma cells are indicative of MGUS. MGUS is often an incidental finding, and while it typically has a benign course, approximately 1% of MGUS cases per year evolve into multiple myeloma or other lymphoproliferative disorders.[8]

The transition from MGUS to multiple myeloma is defined by the increase in plasma cells within the bone marrow, as well as the increase in free light chains and the development of systemic symptoms.[9] In our patient, kappa/lambda ratio imbalance and the increase in plasma cells to 30% on repeat bone marrow biopsy, combined with worsening systemic symptoms, skin ulcers, and neuropathy, pointed towards the progression to multiple myeloma.

Previous studies have reported only a small percentage of pSS patients progressing to overt malignancies, including MM, with some evidence linking the chronic inflammatory state in pSS to such malignant transformations.[10] The presentation of this patient with cutaneous vasculitic lesions, peripheral neuropathy, and transformation of MGUS to MM over a short span of one year adds to the complexity and rarity of this case. Most cases in literature describe MGUS remaining stable or progressing over many years, rather than such a rapid evolution. Additionally, the recurrence of vasculitic skin lesions, neuropathy, and worsening sicca symptoms during the transformation to MM, in conjunction with C4 hypocomplementemia, has not been frequently emphasized in similar cases. This combination of factors highlights the unique nature of this case.

CONCLUSION

This case highlights the rare and rapid transformation of MGUS to multiple myeloma in a patient with pSS. It underscores the importance of vigilant monitoring for paraproteinemia and systemic complications in pSS patients, as early detection and intervention can significantly impact outcomes. The unique presentation of this case, with systemic features like constitutional symptoms, recurrent vasculitic lesions, neuropathy and with risk factors like hypergammaglobulinemia, presence of autoantibodies anti-Ro/SSA and hypocomplementemia, provides valuable insights into the disease evolution and the potential for malignant transformation. Multi-disciplinary management remains essential to address the complex interplay of autoimmune and hematologic processes.

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