MGUS In Patient With Chronic Urticaria: Schnitzler's Syndrome

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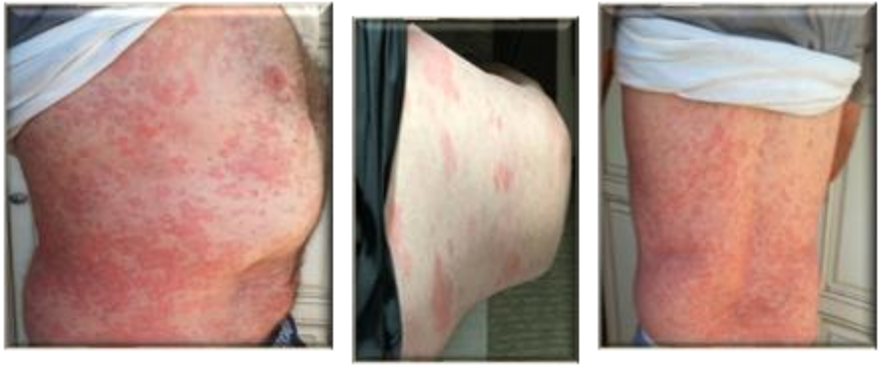
Introduction

- Schnitzler's syndrome is a rare autoinflammatory disease characterized by chronic urticarial hives and elevated monoclonal gammopathy that is associated with malignancy that is underdiagnosed.
- Diagnostic criteria includes presence of monoclonal IgM or IgG immunoglobulin, chronic urticaria, joint pain and elevated inflammatory markers.
- Schnitzler's syndrome is considered a premalignant clonal plasma cell disorder that may lead to potentially life-threatening complications.
- Potential malignant transformation includes multiple myeloma or myeloproliferative disorders.
- Blockade of interleukin-1 (IL-1), a key cytokine in the pathogenesis of the disease, dominates current therapeutic protocols.
- Anakinra (Kineret), a recombinant Human IL-1 receptor antagonist and Cyclosporin is the most widely used treatment options.

Case Description

- We highlight a case of a 75-year-old male with a history of chronic idiopathic urticaria who initially presented with joint pain, urticaria skin lesions located on his torso, back, shoulder, ankles and bilateral inner medial thighs for past seven years.
- Multiple treatment modalities were utilized for his symptoms; however, the urticaria did not improve with H1, H2 blockade, leukotriene antagonist, doxepin, and five doses of Xolair which worsened the urticaria. Corticosteroids helped with urticaria. However, Did not resolve urticarial lesions.
- Evaluated by both allergy, rheumatology and hematolgy.
- Patients' rash resolved with the start of cyclosporin



Snapshot of Patient Workup

Measure	Result
Hematological workup SPEP: Total Protein/Serum Protein electrophoresis UPEP : Urine Protein Electrophoresis Tryptase: Leukocyte and Lymphocyte flow showed decreased B-Cells and no finding malignancy.	SPEP-Elevated IGM Lamda M Spike on SPEP UPEP no M Spike Tryptase is Negative
B2-Microalbumin	4.45 HIGH
Punch Biopsy Skin	Perivascular and interstitial mixed cell infiltrate with eosinophils.
Rheumatologic workup: Factor (RF), CRP, anti-CCP, endomysial Ab, SSA/B Ab. Endomysial, SSA/B Ab, RF, HIV HTLV-1 and 2 Hepatitis panel, RPR and HIV were negative.	Negative Note: Previous ANA , ESR and RF was elevated. On repeat test they normalized.
PET CT	No osseous lesions

Conclusion

- Schnitzler's Syndrome is a rare disease that is underdiagnosed.
- Evaluation of underlying hematologic malignancy or premalignancy should be included in clinician's workup with chronic urticaria.
- 10-15% of Schnitzler's syndrome of patients will eventually develop a lymphoproliferative disorder, such as lymphoplasmacytic lymphoma, or IgM myeloma.
- A missed diagnosis of Schnitzler's syndrome can have serious health consequences for the patients.

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