VIRGINIA DERMATOLOGY SOCIETY

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The Great Mimicker: Systemic Lupus Erythematosus Presentation as Toxic Epidermal Necrolysis

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An 18-year-old female presented to the Emergency Department with an evolving rash. The patient was found to be febrile at 103.5F but hemodynamically stable. Laboratory findings revealed anemia, thrombocytopenia as well as acute liver and renal failure. Physical examination highlighted dusky, erythematous, targetoid vesicles on the face, neck, upper extremities and trunk as well as dusky, erythematous targetoid papules and plaques on the bilateral lower extremities and hemorrhagic crusting of the lips. Given the rapid progression of a Nikolsky positive, desquamating rash with mucosal involvement, a thorough review of systems (ROS), medication history and skin biopsy was performed. While the ROS was non-specific and medication history was unrevealing, the skin biopsy noted full thickness epidermal necrosis, follicular epithelial necrosis and sub-epidermal vesiculation with mild lymphocytic and neutrophilic inflammation. Histologic direct immunofluorescence (DIF) was negative. Steven Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) were in the original differential diagnosis. Further laboratory testing highlighted positive anti-nuclear antibody testing (ANA 1:2560, speckled pattern) and anti-SSA/Ro antibodies. The leading diagnosis for our patient was acute systemic lupus erythematosus (SLE).

Systemic lupus erythematosus presenting as SJS/TEN is an extremely rare entity with most publications highlighting less than 50 cases worldwide. The atypical presentation and a negative DIF pattern resulted in a hesitation to diagnose SLE; however, drug hypersensitivity and infectious etiology were sufficiently ruled out. It is important to recognize that SLE may present as a life threatening rash even in the setting of a negative DIF. The treatment required to temper this proinflammatory state includes systemic glucocorticoid, azathioprine, mycophenolate mofetil and cyclophosphamide. Other treatment modalities include intravenous immunoglobulin (IVIG) therapy and plasmapheresis. In our patient, tumor necrosis factor inhibitor therapy, systemic glucocorticoid, IVIG and mycophenolate mofetil were used to adequately control SLE symptoms in the setting of multi-organ involvement.

References:

Baker, M.G Cresce, N.D. Ameri, M. Martin, A.A et al. “Systemic Lupus Erythematosus Presenting as Steven Johnson Syndrome/Toxic Epidermal Necrolysis.” *Journal of Clinical Rheumatology*. 20:3 (2014), 167-171.

Cetin, G.Y. Sayar, H. Ozkan, F. Kurtulus, S. Kesici, F. Sayarhoglu, M. “A Case of Toxic Epidermal Necrolysis-Like Skin Lesions with Systemic Lupus Erythematosus and Review of the Literature.” *Lupus.* 22 (2013), 839-846.

Lee, H-Y. Tey, H-L. Pang, S-M. Thirumoorthy, T. “Systemic Lupus Erythematosus Presenting as Steven-Johnson Syndrome and Toxic Epidermal Necrolysis.” *Lupus*. 20 (2011), 647-652.

Tankunakorn, J. Sawatwarakul, S. Vachiramon, V. Chanprapaph, K. “Steven-Johnson Syndrome and Toxic Epidermal Necrolysis-Like Lupus Erythematosus.” *Journal of Clinical Rheumatology*. 25:5 (2019), 224-231.

Torchia, D. Romanelli, P. Kerdel, F.A. “Erythema Multiforme and Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis Associated with Lupus Erythematosus.” *Journal of the American Academy of Dermatology*. 67:3 (2011), 417-421.

Zeimer, M. Kardaun, S.H. Liss, Y. Mockenhaput, M. “Steven-Johnson Syndrome and Toxic Epidermal Necrolysis in Patients with Lupus Erythematosus: A Descriptive Study of 17 cases from a National Registry and Review of the Literature.” *British Journal of Dermatology*. 166 (2011), 575-600.