ABSTRACT

Background-

Systemic lupus erythematosus (SLE) is a multisystem, autoimmune disease, involving complex pathogenetic mechanisms that can present at any age. Late onset SLE is a subgroup of SLE defined as SLE diagnosed at or over the age of 50. Angioedema is a self-limited, localized swelling of the skin and mucosal tissues (larynx, bowel wall), which results from extravasation of fluid into the interstitium due to loss of vascular integrity. Angioedema has been rarely associated with SLE due to antibody formation against the C1 esterase enzyme causing activation of the bradykinin pathway.

Case presentation-

58-year-old male presented with fever and pleuritic type chest pain, was found to have pancytopenia, sub nephrotic range proteinuria, positive ANA and dsDNA, and was diagnosed to have SLE. After one month, he again presented with angioedema with low C1 esterase inhibitor functional levels and his edema resolved with steroids. His SLE remained stable without any angioedema up to date with a minimum maintenance prednisone dose.

Conclusion –

To the best of our knowledge this is the first case report which describes angioedema associated with late onset male SLE. This case report highlights the importance of connective tissue disorders to be kept in mind as a rare acquired cause for angioedema, especially when a patient with a background of a diagnosed connective tissue disorder, presents with angioedema without an obvious cause.