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Multiple subcutaneous nodular lesions in a patient with SLE – A rare form of cutaneous lupus – Lupus profundus: A case report

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Background

Lupus erythematosus panniculitis (LEP) also known as lupus profundus, presents with recurrent subcutaneous inflammatory lesions which are classified under chronic cutaneous lupus erythematosus (CLE). It appears as deeply indurated tender plaques or nodules which can cause ulcerations and scaring. It involves the proximal extremities, trunk, face, and scalp leading to disfiguration. Rarity of the disease and wide range of clinical presentations can cause significant diagnostic and treatment delays. We describe a patient with SLE who later developed a generalized nodular lesions due to lupus profundus. She was successfully treated with Hydroxychloroquine (HCQ) and Mycophenolate mofetil (MMF).

Case presentation

A 51 year old female patient who presented with mononeuritis multiplex, a vasculitic rash over right lateral aspect of the knee, constitutional symptoms, malar rash, hair loss, positive ANA and low C3, C4; was diagnosed with systemic lupus erythematosus (SLE). 6 months following the diagnosis she experienced a tender nodular skin lesions first appeared in the abdomen which gradually ulcerated. Subsequently she developed similar nodules in proximal arms and thighs. She also had a biopsy confirmed myositis with CPK over 4000 IU/dL. Initial differentials included dermatomyositis or SLE /dermatomyositis overlap syndrome. Inflammatory markers, ANCA, Ds DNA, U1 RNP and Jo-1 Antibody were all negative. Skin biopsy was reported as panniculitis. The clinical diagnosis of lupus profundus and lupus myositis was made. Since the myositis was severe resulting in dysphagia, she was treated with methyl prednisolone three pulses followed by IV cyclophosphamide fortnightly six pulses. Skin lesions which were resistant to initial treatment, later responded to MMF.

Conclusion

Lupus profundus is a rare clinical entity of cutaneous lupus erythematosus. It can present preceding or following the diagnosis of SLE. Increased awareness on this rare clinical presentation would avoid delays in diagnosis. Early commencement in treatment prevents disfigurement. Antimalarials and other immunosuppressants have been used in the management. However, definitive treatment guidelines are yet to be established.

Key words: Systemic Lupus Erythematosus, Subcutaneous nodular lesions, Lupus profundus