

ACQUIRED ANGIOEDEMA IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Abstract: *Angioedema due to acquired C1-inhibitor (C1-INH) deficiency (also referred to as acquired angioedema) is a rare, life-threatening disease with poorly defined aetiology, therapy, and prognosis. The article reports on acquired angioedema in 8 patients with systemic lupus erythematosus (SLE) representing 16.66% of the total 48 SLE monitored in our clinic. Sex ratio was 1/7, medium age 45.5±12.61 years. In 7 cases angioedema preceded the diagnosis of SLE with 13.28 ±7.8 months, one case developed angioedema 15 years after SLE diagnosis. At the time of acquired angioedema diagnosis the medium C1-INH concentrations were 0.094±0.039 g/L and C4 was decreased in all cases, medium values 14.42± 4.83 mg/dl. Angioedema localization was at face/eyelids in 5(62.5%) cases, larynx in 3(37.5%) cases and 2(25%) cases had severe abdominal pain. 3(37.5%) patients also associated pruritic erythematous rash. Compared with the SLE patients not having angioedema, in the acquired angioedema group there was increased incidence of systemic symptoms, immune cytopenias, kidney involvement, antiRo and anti Sm antibodies. Patients evolved with a medium of angioedema attacks of 6.87/year until SLE was diagnosed; after treatment for SLE was started the frequency decreased to 2.28 attacks/year and entered into angioedema complete remission after 11.5 months. C1-INH concentration reached normal values after 12± 2.16 months in all 7 cases where angioedema preceded SLE diagnosis and remained low in the SLE patient developing angioedema after SLE was diagnosed. Acquired angioedema responded best to SLE treatment with hydroxychloroquine or immunosuppressant*

Key words: *angioedema, systemic lupus erythematosus*

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