SKIN MANIFESTATIONS OF INTERNAL DISEASE

AN UNUSUAL CLINICAL PRESENTATION OF ACQUIRED C1 ESTERASE INHIBITOR DEFICIENCY ANGIOEDEMA

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Background: Acquired C1-esterase inhibitor (C1-INH) deficiency angioedema (C1-INH-AAE) is a form of bradykinin-mediated angioedema. A quantitative or functional C1-INH deficiency with negative family history is diagnostic of C1-INH-AAE The diagnosis should be considered when patients present with isolated angioedema without urticaria in the fourth decade of life or later without a family history of angioedema. Herein, we report an unusual clinical presentation of C1-INH-AAE.

Observation: A 40 –year-old women, without a relevant medical history, presented to our outpatient clinic department with recurrent painless white edema white located at the anterior face of the left elbow. This lesion lasted 3 days. A day later a second lesion appeared on the anterior face of the same forearm. The evolution was marked by increased edema of the anterior and antero-internal lower third of the left forearm with hypoesthesia. This patient had spontaneous pain of the wrist. We have ruled out the diagnosis of thrombophlebitis. The dosage of C1 inhibitor was normal but the dosage of its activity was low. The radiological and biological explorations did not show any abnormality especially hematological malignancies.

Key message: Angioedema acquired by functional deficit in C1 inhibitor is a rare pathology. The presentation of our patient with lesions in forearm is unusual. The long-term prognosis is determined by associated hematologic malignancies.