Anti- β_2 -Glycoprotein I Antibodies in Sneddon's Syndrome

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Sneddon's syndrome (SNS) consists of generalized broken livedo reticularis and ischemic cerebral manifestations [1, 2]. Its relationship with systemic lupus erythematosus (SLE) or antiphospholipid syndrome is a matter of discussion, since clinical and/or biological markers of SLE and/or APS are inconstantly found in patients with SNS [2].

It has been shown that some anticardiolipin (ACL) antibodies require the presence of a plasma protein, β_2 -glycoprotein I (β_2 gpl), for binding to their target [3, 4]. An ELISA technique using purified human β_2 gpl (Dr. H. Haupt, Behringwerke, Marburg, FRG) has been recently developed for the detection of anti- β_2 gpl antibodies [5]. A pilot study in 47 patients with SLE suggests that anti- β_2 gpl antibodies are strongly associated with thrombotic events in those patients [5].

We looked for the presence of anti- β_2 gpl antibodies in 16 consecutive patients with SNS.

Patients included 14 women and 2 men. The mean age at onset of cerebrovascular disorder was 37 years (22-58). Livedo was already present from 2 to 36 years at onset of neurological events. Four of the 16 patients had SLE according to ARA criteria [6]. Lupus anticoagulant was detected by coag-

ulation tests in 5 patients (including 2 with SLE). ACL antibodies were detected by ELISA test in 7 (including 3 with SLE). Nine patients had neither lupus anticoagulant nor ACL antibodies (including 1 with SLE).

The method for detection of anti- β_2 gpl antibodies is described elsewhere [5]. Results above mean $\pm$ 3 SD of normal controls were considered as positive. Among the 16 patients with SNS, 11 (68.7%) had anti- β_2 gpl antibodies. Anti- β_2 gpl antibodies were present in the 4 patients with SLE and in 5 of the 9 patients who had neither lupus anticoagulant nor ACL antibodies.

These preliminary data suggest that anti- β_2 gpl antibodies are a new biological marker of patients with SNS, more frequently encountered than lupus anticoagulant or ACL antibodies. Further studies, including serial determinations, are needed to determine its exact prevalence in SNS.

The speculative hypothesis that the presence of anti- β_2 gpl antibodies could identify among CNS symptom-free patients with generalized broken livedo reticularis those who are prone to develop cerebrovascular events should be tested prospectively.

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