INTERNAL MEDICINE

□ EDITORIAL □

Adrenal Insufficiency Complicated with Antiphospholipid Syndrome

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Key words: adrenal insufficiency, antiphospholipid syndrome

(DOI: 10.2169/internalmedicine.45.0167)

Antiphospholipid syndrome is characterized by the clinical evidence of vascular thrombosis, recurrent fetal loss, and thrombocytopenia accompanied by an elevated level of various kinds of antiphospholipid antibodies (1). The syndrome is either primary or secondary to an underlying condition, most commonly systemic lupus erythematosus (2). We previously reported overt congestive heart failure with mitral and aortic regurgitation due to antiphospholipid syndrome in a patient with systemic lupus eryhthematosus (3). The spectrum of clinical manifestations appearing in patients with antiphospholipid syndrome is wide and diverse, including neurological disorders such as chorea and cutaneous features such as livedo reticularis (1). Vascular abnormalities have also become more recognized as one of the important features of this syndrome, although such abnormalities are clinically silent in most of cases (4-10).

The adrenal manifestations of the antiphospholipid syndrome have received little attention or are completely omitted in large series (11, 12). In patients with primary antiphospholipid syndrome, spontaneous bilateral adrenal hemorrhage is an uncommon condition that may lead to acute adrenal insufficiency and death. It was almost exclusively a postmortem diagnosis before computed tomography scan was developed. Anecdotal cases of survival have been reported in patients with bilateral adrenal hemorrhage associated with the antiphospholipid syndrome (13). The diagnosis of adrenal insufficiency in patients with bilateral adrenal hemorrhage was rarely made before death 10 years ago, because the sign and symptoms are not dramatic and were nonspecific (14). After an abdominal CT scan showing a bilateral enlargement of the adrenal glands, a presumptive treatment should be started while confirmation of adrenal insufficiency is made by hormonal evaluation; low basal cortisol and aldosterone levels, and the absence of rise in plasma cortisol after a short corticotrophin stimulation test are the most commonly used tools for the diagnosis of adrenal insufficiency (15).

Recently, 20 patients with acute adrenal failure as the heralding symptom of primary antiphosphlipid syndrome were reviewed (16). The great majority of them were males (75%) with a mean age of 42 years. Abdominal pain was present in 14 patients, followed by fever (13 patients) and hypotension (12 patients). The main morphological findings by computed tomography or magnetic resonance were committed with bilateral adrenal hemorrhage in 11 patients. Lupus anticoagulant was positive in all of 19 patients. Recently, Fujisawa et al reported a typical patient with adrenal insufficiency complicated with antiphospholipid syndrome indicating hyponatremia, hypoglycemia and prolongation of activated partial thromboplastin time in laboratory data and bilateral adrenal enlargement by computed tomography (17).

On the basis of the above cases it is important to rapidly diagnose adrenal insufficiency and assess clotting time. Antiphopsholipid syndrome should be considered when we encounter patients with bilateral adrenal hemorrhage and symptom of adrenal insufficiency.

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