Acute adrenal failure secondary to bilateral infarction of the adrenal glands as the first manifestation of primary antiphospholipid antibody syndrome

Adrenal insufficiency is an uncommon, life threatening complication of the primary antiphospholipid antibody syndrome (PAPS), secondary to either adrenal haemorrhage or infarction. Adrenal failure more often follows other PAPS thromboembolic manifestations, and has to be clearly differentiated from other PAPS abdominal emergencies, including mesenteric and hepatic infarction. The pathological mechanisms involved are still not clearly understood, but the hypercoagulable state in patients with PAPS supports the concept that adrenal haemorrhagic infarction may possibly be related to adrenal vein thrombosis. We recently observed a new case, which is of particular interest as the patient developed an acute adrenal failure, revealing bilateral adrenal infarction, as a first manifestation of the PAPS.

A 70 year old patient presented with diffuse abdominal pain persisting for 15 days. He had no previous history of thromboembolic or connective tissue disease. On admission, he was dehydrated with pulse 120/min, blood pressure 9/4 mm Hg, and abdomen palpation was tender. Laboratory findings were: haemoglobin 6.3 mmol/l, white cell count 6.4 ×10⁹/l, platelets were 90 ×10⁹/l, ESR 18 mm 1st h. Blood electrolytes were abnormal for natraemia 118 mmol/l, kaliaemia 6.3 mmol/l, glycaemia 2 mmol/l. Acute adrenal failure diagnosis was suspected, and was confirmed by a low plasma cortisol concentration 20 nmol/l and an adrenocorticotrophichormone (ACTH) test that failed to raise plasma cortisol over 25 nmol/l. The serum ACTH concentrations were 300 pg/l (normal range 10–50). Coagulation studies found a pronounced prolonged activated partial thromboplastin time (APTT) (95 s; normal: 32 s) with normal activated partial thromboplastin time inhibition test with a high dilution of thromboplastin (1:500); the confirmatory test for LAC was positive. Anticardiolipin antibodies IgG were positive (IgG-aCL: 18 GPL U/ml; normal < 13) and IgM were negative. The presence of aCL was determined by solid phase enzyme linked immunosorbent assay according to international standardised methods. Examination of antiphospholipid antibodies (asearchrom APA-Diagnostica Stago) was negative. The results of autoantibody screening, including Treponema pallidum haemagglutination, Venereal Disease Experimental Laboratory test, antinuclear and anti-DNA antibodies, cryoglobulin, and rheumatoid factor were negative. The complement profile was normal. Abdominal computed tomography showed bilateral enlarged adrenal glands, secondary to adrenal haemorrhagic infarction (fig 1). The diagnosis of PAPS was made, because of the presence of LAC and IgG-aCL, associated with bilateral adrenal infarction, which was probably secondary to thrombosis of the adrenal gland veins. The patient was treated with cortisone acetate and aspirin, with rapid improvement of his clinical status. Three months later, and while APTT, IgG-aCL, and LAC titres were still raised, the patient developed an extensive deep venous thrombosis of the right forearm, confirmed by venous Doppler echography. Anticoagulation treatment was begun.

The PAPS is characterised by recurrent venous or arterial thrombosis, or both, or repeated fetal loss, associated with the persistent presence of anticardiolipin antibodies or LAC, in the absence of connective tissue disease (notably systemic lupus erythematosus). Our case report is original in that the acute adrenal failure, secondary to bilateral adrenal haemorrhagic infarction, was the first clinical manifestation of a typical PAPS. We suggest therefore, that PAPS may be suspected in patients with either acute or chronic adrenal failure, even if they have no previous history of thromboembolic disorders. Because haemorrhagic infarction may precede other thromboembolic events, when this type of complication is noted, an evaluation for PAPS with a search for antiphospholipid antibodies should be systematically done. Moreover, adrenal haemorrhagic infarction secondary to PAPS should be excluded in all patients presenting with enlarged adrenal glands shown by abdominal computed tomography. Our findings further emphasise that diagnosis of adrenal insufficiency should be considered in patients with PAPS and acute abdominal pain.

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Echocardiographic findings in primary Sjögren’s syndrome

Primary Sjögren’s syndrome (pSS) is a chronic autoimmune disease characterised by lymphocytic infiltration of the salivary and lacrimal glands.1 Similar lymphocytic infiltration can be observed in visceral organs, and this results in several extraglandular manifestations.2 Among these, a clinically overt heart disease is very rare.3 However, recent echocardiographic studies showed that asymptomatic cardiac involvement is frequent in pSS. Thus, Rantaapää-Dahlqvist and colleagues4 reported unexpected high frequency of cardiac manifestations in primary Sjögren’s syndrome: imaging findings of pericardial effusion were seen in 21.2% of 18 female patients diagnosed with Sjögren’s syndrome, including 9 patients with clinical or subclinical pericardial effusion which was confirmed by echocardiography. They reported abnormal findings in three patients, respectively. No patient had history of cardiovascular diseases, such as arterial hypertension or ischaemic heart disease. Echocardiography was carried out with an ATL Aposcope 800 instrument and normal values of the measured parameters were taken from Feigenbaum.5 Transmural diastolic flow velocities were recorded by pulsatile Doppler method. Moreover, the left ventricular diastolic function was evaluated according to Cochrane.6 Statistical analysis was performed using the Student’s t test and Scheffe’s method for multiple comparison among means. The results show that only the deceleration of the E wave was significantly reduced in pSS (mean SD) (360 (84.02) cm/s)2 compared with controls (462 (84.25) cm/s)2 (p<0.0009), and remained significantly different when five subjects older than 60 years were excluded from the pSS group. No significant valvular disease was found in both groups. Additionally, present or previous pericarditis and pulmonary hypertension were not detected in pSS.

In conclusion, although overt heart involvement in pSS is very rare echocardiography shows an unexpectedly high frequency of cardiac manifestations, mainly pericarditis and diastolic dysfunction. These findings suggest that cardiac involvement must be included in the spectrum of extraglandular manifestations of pSS.

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