Case report

Catastrophic antiphospholipid syndrome in cancer

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Abstract—Antiphospholipid syndrome is characterized by the presence of antiphospholipid antibodies resulting in arterial and venous thromboembolism. Apart from primary cases, this syndrome is often associated with autoimmune diseases. Around 50 cases of catastrophic antiphospholipid antibody syndrome have been reported as yet. Authors describe the first case of catastrophic antiphospholipid syndrome associated with gastric cancer. Apart from presenting the clinical case, authors also discuss the possible pathomechanism of this associated disorder including the role of immunological factors, as well as antiphospholipid antibodies.

Key words: Antiphospholipid syndrome; anti-cardiolipin antibodies; β2 glycoprotein I; gastric cancer.

INTRODUCTION

Antiphospholipid (APL) antibodies can be detected in 1–2% of healthy individuals but their existence is more common in autoimmune diseases [1]. They include antibodies to cardiolipin and to its cofactor β2 glycoprotein I, as well as lupus anticoagulant. In the presence of these antibodies, there is a higher prevalence of arterial and venous thrombotic events and spontaneous abortions [1–5]. Antiphospholipid syndrome (APS), is associated with APL antibodies [6]. The diagnosis of APS is based on the detection of APL antibodies, as well as on the coexistence of arterial and/or venous thrombotic events, recurrent abortions and thrombocytopenia. Recently, an international consensus statement on the preliminary classification criteria for APS was published [7]. These criteria are based on clinical signs includ-
ing thrombotic events and fetal losses, as well as laboratory markers, such as anti-cardiolipin antibodies and lupus anticoagulant. APS with no association to other immunopathological disorders has been termed primary APS, while APS linked to other autoimmune diseases is known as secondary APS [5, 6, 8].

Catastrophic Antiphospholipid Antibody Syndrome (CAPS) is a severe variant of APS, which becomes fatal in about 60% of the cases. CAPS is a rapidly progressive disorder associated with disseminated thromboembolism in several organs [5, 8–11]. CAPS was first described by Asherson [8]. According to meta-analyses performed on a total of 50 CAPS cases, the mean age of the patients was 26, and CAPS affected 2.4-times more women than men [9, 12]. These patients had high serum concentrations of antibody to β2 glycoprotein I [13]. The disease involves most internal organs including, most commonly, the lungs, brain, skin and kidneys. In CAPS, three or more organs must be affected by APS [8]. CAPS was described in primary form in about 60% of the cases, while in one-third of the patients CAPS was associated with other autoimmune diseases, most commonly to systemic lupus erythematosus (SLE), but some secondary CAPS cases in systemic sclerosis (Ssc) or rheumatoid arthritis (RA) were also published [5, 9, 14]. Until now all reported secondary CAPS cases were associated with autoimmune diseases.

APL antibodies have been detected in the sera of a number of cancer patients. Thrombotic events occur relatively frequently in malignancies [3, 15]. Yet, to our best knowledge, the association of CAPS with cancer has not been reported. Here we present a case of gastric cancer leading to CAPS in a female patient.

**CASE REPORT**

V. M., a 45-year-old female patient had hypertension in her medical history. Prior to admittance, she had had no other illnesses, miscarriages or thrombotic events. It is important to note that her daughter had died with SLE at the age of 22. The patient was admitted to our department in May 1998 with fatigue, weight loss, dysarthria, hemiparesis and epigastrial pain. Clinical examination revealed jaundice and hepatomegaly. However, neuropsychiatric symptoms were dominant including, impaired psychomotility, dysarthria, dysphasia, horizontal nystagmus upon provocation, positive Babinski sign on both sides, and left-side hemiparesis. Upon admittance laboratory tests were performed with the following results: ESR: 12 mm/h, white blood cell count: 16 000 per μl, platelet count: 186 000 per μl, hemoglobin: 122 g/l, hematocrit: 0.36. Differential blood count showed normal distribution of white cells. No fragmentocytes could be observed. Serum bilirubin (23 μmol/l), alkaline phosphatase (1829 U/l), γGT (1000 U/l), ALT (140 U/l), AST (91 U/l), LDH (710 U/l) levels were elevated, while creatinin (75 μmol/l) and urea (3.9 mmol/l) were in the normal range. Serum ammonia was 2-times elevated above the control (70 μmol/l). Markers of hemostasis including partial thromboplastin time and thrombin time were in the normal range, and no signs for