LETTER TO THE EDITOR
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Recurrent Thrombotic Events after Catastrophic Antiphopholipid Syndrome

To the Editor,

A 40-year-old man was transferred to our hospital in March 1994 for gross hematuria and decreased urine output. The patient exhibited chest pain and shortness of breath 2 weeks prior to the initial visit. These symptoms were relieved without particular management. Fever and chills developed thereafter followed by abdominal pain. The patient visited a local clinic and was managed for acute pancreatitis and gastritis for 1 week. However, his symptoms did not subside and urine output decreased progressively.

On admission, he complained of severe headache, nausea, and both flank and back pain. Initial hemoglobin levels and platelet counts were 16.9 g/dL and 478 × 10^9/L, respectively. In the course of 3 days, anemia and thrombocytopenia rapidly developed, with a hemoglobin level of 12.0 g/dL and a platelet count of 29 × 10^9/L. Peripheral blood smear showed schistocytes suggesting hemolytic anemia. Initial blood urea nitrogen and creatinine levels were 69 and 6.0 mg/dL, respectively. Magnetic resonance imaging of the kidney revealed acute cortical necrosis. He also had blood tinged sputum, and the lung perfusion scan revealed perfusion defects in the right lower lobe, suggesting pulmonary embolism. Despite fluid therapy and the administration of diuretics, azotemia progressed rapidly over a 3-day period and he consequently underwent hemodialysis. Azotemia improved with increased urine output only after hemodialysis (Fig. 1).

The serologic results showed the presence of lupus anticoagulant and IgM anticardiolipin antibody, and the patient was diagnosed with antiphospholipid syndrome (APS) with multiorgan failure. Retrospectively, his clinical presentation fits into the category of probable catastrophic APS (CAPS). However, neither intravenous heparin nor warfarin was used due to hemoptysis.

Thereafter, he was admitted several times for coronary events and pulmonary infarction. Two months after the first attack, he was readmitted due to chest pain. The echocardiogram showed inferior and septal myocardial infarction and he was discharged with aspirin and clopidogrel. Three months later, he was readmitted due to chest pain, and a coronary angiogram showed right coronary artery focal stenosis (Fig. 2). He was lost to follow-up for 3 years and was readmitted to the hospital in 1997 for pleuritic chest pain. Chest computed

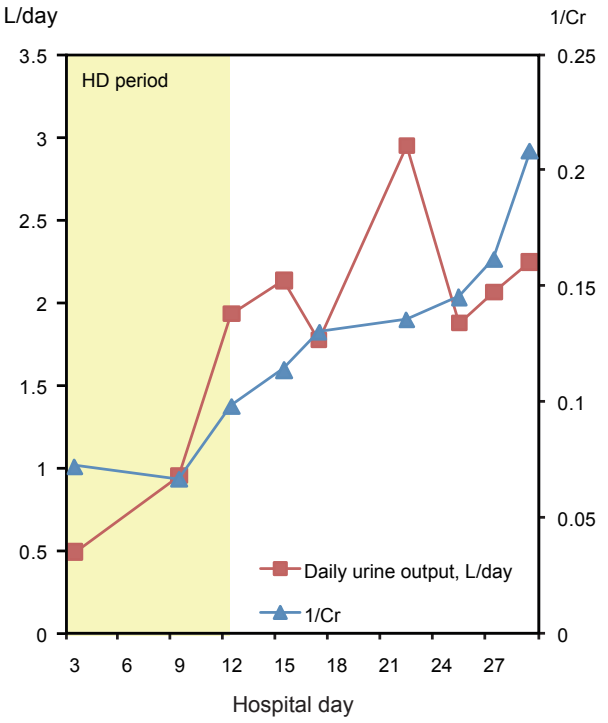

Figure 1. Azotemia progressed rapidly with fluid and diuretics therapy. Urine output increased only after hemodialysis. HD, hemodialysis; Cr, creatinine.

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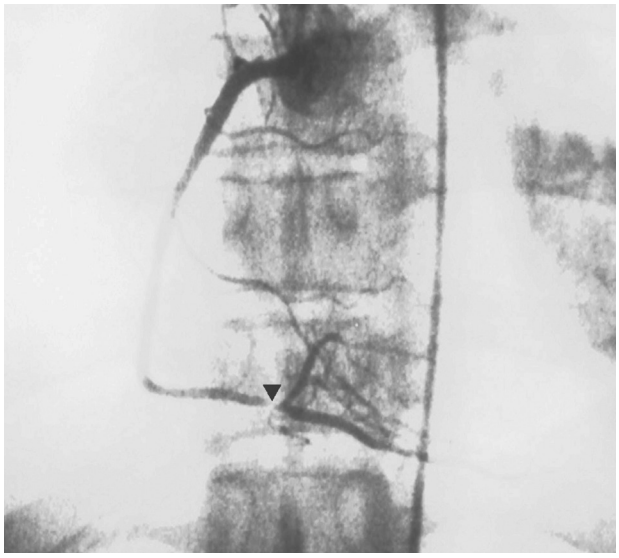

Figure 2. Coronary angiogram showed right coronary artery 50% focal stenosis, suggesting embolic infarct rather than atherosclerosis.

tomography scans revealed both lower lobe pulmonary infarctions. He was lost again to follow-up without antithrombotic agents.

Recently, he was admitted for facial cellulitis associated with a burn, and the involved skin showed multiple ulcerations and necrosis (Fig. 3). His facial lesions improved with antibiotics. We started antithrombotic treatment for long-term prevention of recurrent thrombotic events.

CAPS is a life-threatening medical situation with a high mortality rate, although it represents in less than 1% of patients with APS [1]. Therefore, clinical suspicion and early diagnosis are important for improving survival from CAPS. Asherson et al. [1] and Erkan [2] noted that the recurrence of CAPS was not common, although patients had been on continuous anticoagulation therapy. Among 73 CAPS survivors, 14 patients (19%) had further APS-related manifestations but no patient developed recurrent CAPS episodes during a follow-up of median 67.2 months [3]. Our case presents recurrent APS-related events after an initial CAPS episode.

No standardized treatment guideline for CAPS has been established due to the rarity of the disease and the lack of controlled studies [2]. Current treatment recommendations are based on case series. Previous studies emphasized the importance of early anticoagulation, steroid, and plasma exchange during CAPS episodes [1,4,5]. Bucciarelli et al. [4] demonstrated that the mortality rate decreased from 53% to 33% with a combination therapy of anticoagulation, steroids, and plasma exchange and/or intravenous immunoglobulins in 250 patients in the CAPS registry. The reported patient could not maintain anticoagulation therapy because he had a bleeding episode at the initial admission and did not regularly visit clinics. The recurrent thrombotic events may have been due to the lack of anticoagulation therapy. This case suggests that continuous anticoagulation is important for the prevention of recurrent thrombotic events after CAPS.

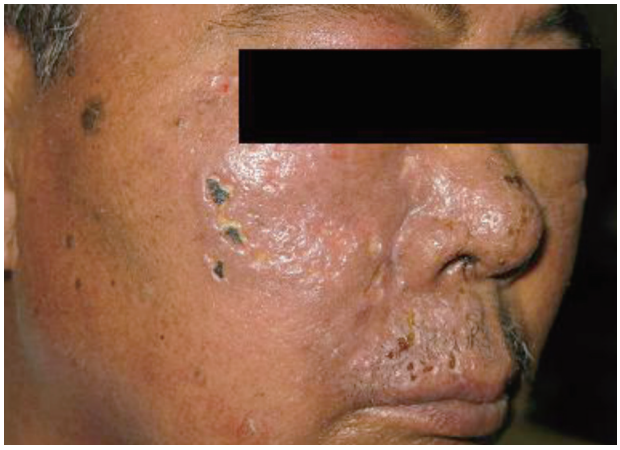

Figure 3. The patient developed facial cellulitis with multiple necrotic ulcerations after burn, suggesting possible microangiopathic thrombosis.

Keywords: Antiphospholipid syndrome; Thrombosis; Anticoagulants

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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