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Therapeutic plasma exchange in catastrophic antiphospholipid syndrome: a rare case with concomitant systemic lupus erythematosus and infection

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A 49-year-old female patient with a 16-year history of systemic lupus erythematosus (SLE), previously without signs of renal involvement (antinuclear antibodies in titer 1: >20480 identified with immunoblot method as anti-histones, anti-nucleosomes, anti-SSB, and anti-RNP) in remission for over 6 months, was admitted to the Immunology Department at University Hospital in Krakow. She had been receiving chronic but irregular treatment with systemic steroids due to poor compliance. At the time of admission, the patient was receiving treatment with 10 mg of prednisone daily. The admission was prompted by the presence of fever, digital ulcers and erythematous-blistering lesions with painful skin necrosis on the abdomen and lower chest (Figures 1A, 1B). Further examination revealed hypotension not requiring the use of pressor agents, and decreased vesicular murmur at the lung bases. The

laboratory test results revealed anemia (hemoglobin 7.8 g/dL, reference range: 11.20–15.70 g/dL), thrombocytopenia (38000/uL, reference range: 180000–370000/uL), prolonged prothrombin time (18.5 s, reference range: 9.8–12.1 s) and aPTT (118.7 s, reference range: 25–33.5 s), decreased eGFR (31 mL/min/1.73m^2, reference range: >90 mL/min/1.73m^2), elevated CRP (465.2 mg/L, reference range: 0–10 mg/L), procalcitonin (37.03 ng/mL, reference range: <0.25 ng/mL), fibrinogen (6.3 g/L, reference range: 1.8–3.5 g/L), d-dimers (7692.6 ng/mL, reference range: <500 ng/mL), and factor VIII (542.2%, reference range: 50–150%). Laboratory tests changes in course of the treatment are presented in Supplementary material, *Table S1*. During hospitalization, the patient experienced severe acute respiratory and kidney injuries, without the need for kidney replacement therapy. A computed tomography scan revealed diffused alveolar hemorrhage (Figure 1C). No thromboembolic signs were detected in the deep vein ultrasound of the lower extremities, as well as in the chest and abdomen computed tomography scans. Based on the clinical findings, positive lupus anticoagulant and IgG anti-cardiolipin antibodies, catastrophic antiphospholipid syndrome (CAPS) was diagnosed. The patient was transferred to the Intensive Care Unit, where invasive mechanical ventilation (IMV) was initiated. Five sessions of plasmapheresis were performed, and pulse of methylprednisolone (500 mg) and rituximab (500 mg) were administered. For the next 4 weeks, the methylprednisolone dose of 1 mg/kg body weight was administered, and then gradually tapered off. Broad-spectrum antibiotic therapy was also initiated at that time (meropenem, linezolid); as the diffused alveolar hemorrhage was stopped, the anticoagulation was continued. The patient's condition transiently improved, but later required reinitiation of IMV, along with the introduction of noradrenaline. There was also a sudden cardiac arrest, successfully resuscitated. A bronchoscopy was performed, along with microbiological examinations of urine and swabs from the secretion of the necrotic skin area. Candida albicans and Acinetobacter baumannii were isolated, and targeted treatment

was initiated (colistin and fluconazole). Following the administered treatment, the patient's condition improved, and she underwent multiple surgical interventions, including removal of the necrotic skin area (Figures 1D, 1E). Towards the end of hospitalization, signs of bone marrow suppression (WBC 2440/uL, reference range: 4000–10000/uL; RBC 2.71 mln/uL, reference range: 3.90–5.20 mln/uL, PLT 142000/uL) were observed, which was confirmed in bone marrow biopsy. However, eGFR, C-reactive protein, and procalcitonin were in normal range (176 mL/min/1.73m2, 5.9 mg/L, and 0.14 ng/mL, respectively); (Supplementary material, *Table S1*). Consequently, the methylprednisolone dose was increased to 1 mg/kg body weight, and a single intravenous dose of immunoglobulins (IVIGs, 1g/kg) was administered, observing a favorable clinical response. Despite the critical condition associated with CAPS likely triggered by infection, and the absence of anticoagulation despite previously detected antiphospholipid antibodies and SLE, the patient was able to return to a state of health stabilization thanks to the collective efforts of the medical team and the broad-spectrum treatment applied, which included antibiotics, anticoagulant medications, immunosuppressive drugs, and plasmapheresis. The patient was discharged home in stable condition and was scheduled for regular monthly follow-ups at the outpatient clinic. The patient passed away 5 months after hospital discharge due to complications of anorexia nervosa.

CAPS is a life-threatening condition characterized by the development of severe thrombotic episodes in a short period, constituting approximately 1% of cases in patients with antiphospholipid syndrome [1]. In terms of clinical manifestations, the kidneys are the most frequently affected organ system (~74%), followed by the lungs (~60%), brain (~56%), heart (~53%), and skin (~45%) [2, 3]. Trigger factors for CAPS are found in approximately two-thirds of individuals, including infections, surgeries, malignancies, ineffective anticoagulation, and SLE flares [4]. The treatment includes anticoagulation and

immunosuppressive drugs (i.e., systemic steroids, cyclophosphamide, rituximab, IVIGs) [1, 2]. The mortality rate of CAPS is around 50%; however, concomitant SLE is associated with a higher mortality rate [5].

Supplementary material Supplementary material is available at www.mp.pl/paim.

Article information

Conflict of interest None declared.

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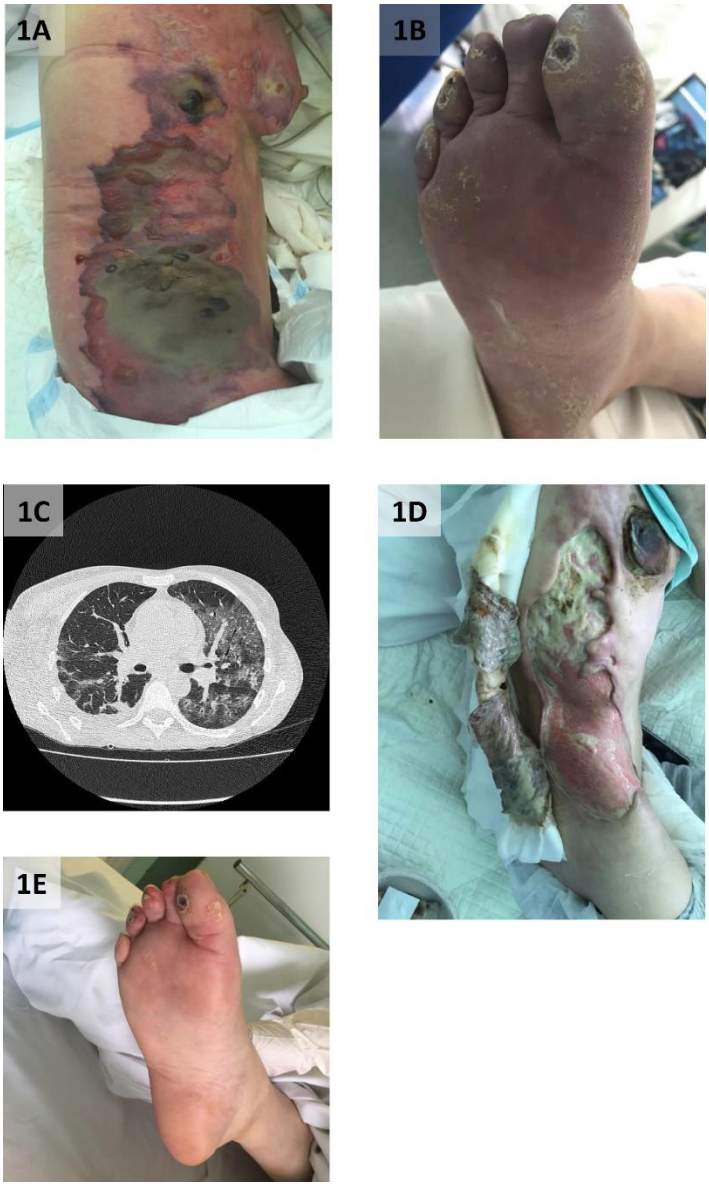

Figure 1 A – Erythematous-blistering lesions with necrosis on the right side of the abdomen and lower chest; B – digital ulcers of the right foot at admission; C – alveolar hemorrhage on

computed tomography scan; D – abdomen and lower chest after removal of the entire necrotic skin area; E – digital ulcers of the right foot after treatment

Short title: Plasma exchange in CAPS concomitant with SLE and infection