

Catastrophic antiphospholipid lung syndrome in young female with repeated abortions

Sir,

Antiphospholipid syndrome (APS) may present with various lung manifestations. We report a case of a 30-year-old female admitted with clinical and laboratory findings consistent with catastrophic APS (CAPS) successfully treated with steroids.

A 30-year-old female presented to the emergency department with a 2-day complaint of acute onset dyspnea, cough with expectoration of blood-tinged sputum, and no fever. She had orthopnea for 2 months but no paroxysmal nocturnal dyspnea chest pain, palpitations, or dizziness. She was married, had three children and gave history 9 abortions.

On examination, she was distressed, pulse 95 beats/min, blood pressure 150/100 mmHg, respiratory rate 30 breaths/min, and temperature 37.5°C with central cyanosis. There had minimal pitting edema bilaterally. Cardiac examination was normal and chest examination only revealed fine late inspiratory crackles on the back.

Chest X-ray (CXR) showed that semi-homogeneous opacities occupy the lower and middle zones of both lung fields and cardiomegaly [Figure 1]; electrocardiogram showed no abnormality but being low voltage. Given the history of the repeated abortions, the acute symptoms with the absence of fever, computed tomography chest with pulmonary angiography was done. The scan showed patches of extensive consolidation involving even the upper lobes [Figure 2] and no embolism in the pulmonary arteries or segmental or subsegmental branches

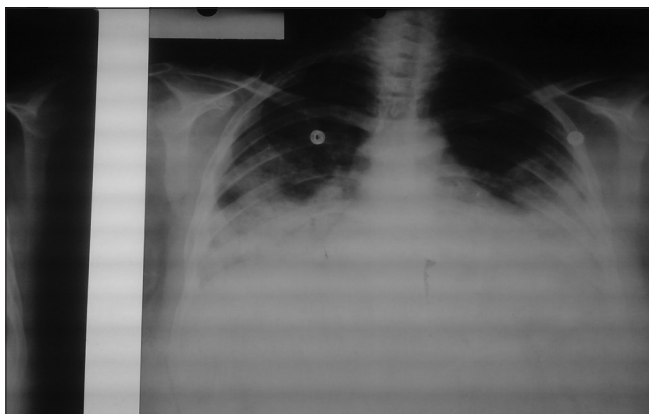


Figure 1: Chest X-ray (posteroanterior view) at admission

and cardiomegaly [Figure 3]. Next day, her respiratory rate increased to 45 breaths/min and arterial blood gas (ABG) analysis showed a pH 7.48, PaCO₂ 28 mmHg, PaO₂ 55 mmHg, HCO₃ 20 mmol/L, and SaO₂ 91%, so the patient was admitted to the respiratory intensive care unit (RICU) for noninvasive ventilation.

In the RICU, the patient was empirically treated with broad-spectrum antibiotics along with noninvasive ventilation. Routine laboratory investigations were done. The C-reactive protein was elevated (102 mg/L), and the erythrocyte sedimentation rate was 61/85. The complete blood count showed white blood cells 18.000/mm³, red blood cells 4 million/mcL, hemoglobin 10 g/dl, and platelets 177.000/mm³. Renal and liver functions, serum electrolyte, and coagulation profile were within normal ranges. Echocardiography assessment was done which detected increased diameter of the left ventricle and impaired contractility. The antinuclear antibodies and anti-DNA were requested. Treatment with prednisolone 60 mg/day was started along with the previously mentioned medications.

After 2 days, the patient reported improvement in her symptoms. The respiratory rate decreased to 24 breaths/min, and ABG on venture mask 35% was as follows: pH 7.44, PaCO₂ 32 mmHg PaO₂ 67 mmHg, HCO₃ 23 mmol/L, and SaO₂ 90%. CXR follow-up [Figure 4] showed some improvement. Detailed echocardiography was done and revealed global hypokinesia with segmental variation (ejection fraction 44%), dilated left atrium, and left ventricle and moderate mitral regurgitation. Urine analysis showed proteinuria (+2) and 24-h urinary protein level was increased (2198 mg/24 h, normal up to 150 mg/24 h). Lupus anticoagulant (LA), anticardiolipin (aCL) immunoglobulin G, and immunoglobulin M were done and were positive. The patient was diagnosed with systemic lupus erythematosus (SLE) and secondary antiphospholipid antibody (aPL) syndrome. She received hydroquinone 200 mg twice per day, aspirin, and warfarin. Steroids were gradually withdrawn. Before discharge, follow-up CXR showed marked improvement [Figure 5] and so did the ABG analysis (on room air, the patient achieved pH 7.46, PaCO₂ 35 mmHg, PaO₂ 70 mmHg, HCO₃ 24 mmol/L, and SaO₂ 97%). The patient was discharged and advised to follow up with a rheumatologist.

The CAPS is a fatal variant of APS, with a prevalence of 1% of APS population. It was first described in 1992 and

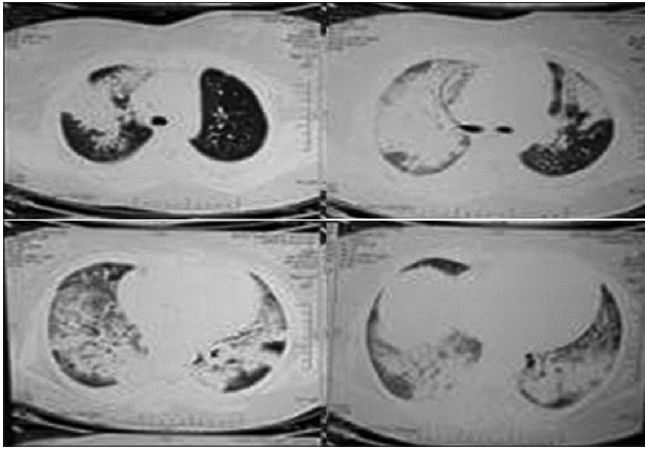


Figure 2: Multislice computed tomography chest on admission (pulmonary window)

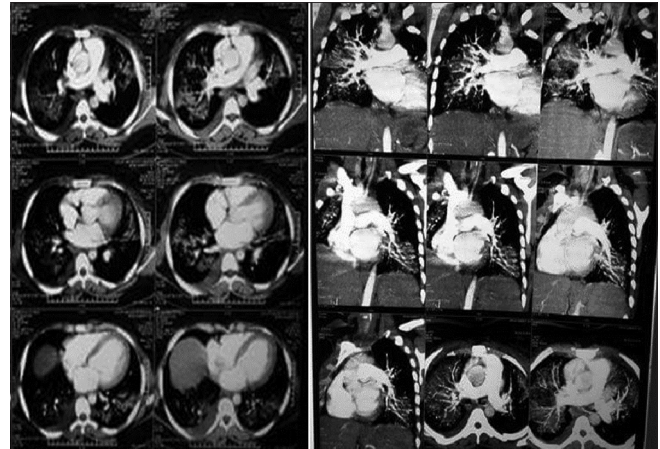


Figure 3: Multislice computed tomography angiography on admission

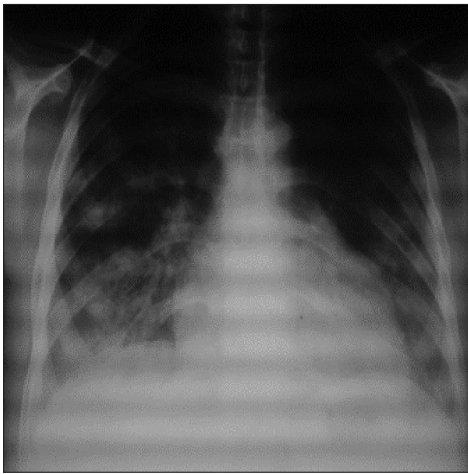


Figure 4: Chest X-ray follow-up after 2 days of steroid

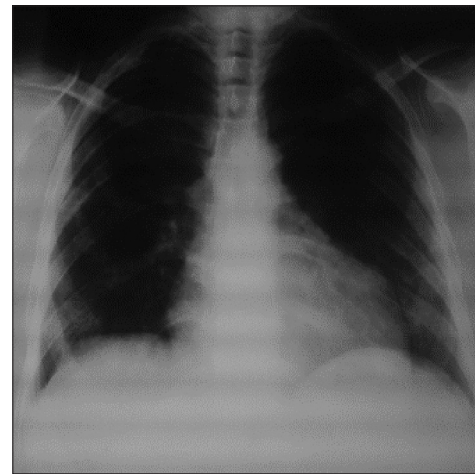


Figure 5: Chest X-ray follow-up at discharge

defined as thrombosis of at least three different organ systems with histopathologic evidence of multiple small vessel occlusions and high titers of aPLs.^[1-5]

As regard pathogenesis of CAPS, several mechanisms have been suggested including infections, molecular mimicry, activation of endothelium microvasculature, and small vessel occlusions which have led to systemic inflammatory response syndrome and release of inflammatory cytokines, complement (C3, C5) which in combination with aPL antibodies resulting in the characteristic thrombosis of CAPS.^[3,6] In Catastrophic Antiphospholipid Syndrome (CAP) study analysis, gender was females in 72%. As the majority of cases suffered from primary APS (40%), SLE, lupus-like disease, and other autoimmune diseases. Interestingly, CAPS was the first manifestation of APS in 46% of the 280 patients. A detailed description of the clinical manifestations^[7] and treatment^[8] of CAPS can be found elsewhere.

The clinical manifestation of CAPS depends on organ affected by thrombosis including kidney (71%), lung (64%), brain (62%), heart (51%), and skin (50%).^[7-9]

Our patient presented with acute respiratory distress syndrome, hypertension, left ventricular dysfunction, acute renal injury, and high titer of aPLs and history of repeated abortions. It has been reported that hypertension in CAPS tends to occur in association with renal involvement.^[5] Since 1998, in the study of 50 patients with CAPS, it was reported that 78% of patients had renal involvement associated by hypertension which was often malignant. Renal biopsy shows thrombotic microangiopathy which is a pathologic hallmark of CAPS.^[4,10] Laboratory findings in CAPS patients may include thrombocytopenia, hemolytic anemia which is often accompanied by schistocytes, and disseminated intravascular coagulations (DIC).^[6] The autoantibodies which used for the diagnosis of APS are anti-B2-glycoprotein I, aCL, or LA assay.^[1,9]

Differential diagnosis for CAPS includes sepsis, hemolytic uremic syndrome, thrombotic thrombocytopenic purpura, and DIC.^[11] The differential diagnosis for CAPs from other microangiopathic syndromes may be challenging as the acute onset of thrombosis leading to multiple organ failure; hence, early diagnosis and management are very important for patient survival.

Treatment guidelines for CAPS include a combination of corticosteroids, anticoagulants, intravenous immunoglobulins, plasma exchange, and cyclophosphamide.^[6,12] However, new therapeutic modalities have been used in refractory CAPS including rituximab, defibrotide, and eculizumab.^[13]

Taken together, CAPS is a rare but potentially life-threatening variant of APS. Early diagnosis and management are imperative for patient survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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