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Catastrophic antiphospholipid syndrome (CAPS) – A case report

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Case report

Catastrophic antiphospholipid syndrome (CAPS) – A case report

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Abstract

Catastrophic antiphospholipid syndrome is a rare disease with positive antiphospholipid antibodies that progresses to a life-threatening condition resulting in multiple organ dysfunction and thromboses. Here is a 19-year young woman who presented to the emergency room in a haemodynamically unstable condition requiring immediate mechanical ventilator support. She had involvement of the disease in the respiratory, cardiovascular, renal, and central nervous systems. She also developed gangrene of the left toes for which amputation was done. She was successfully treated with Dexamethasone Cyclophosphamide Pulse (DCP) therapy. The collaborative care by the health team members in the intensive care unit was successful in preventing complications and enhancing speedy recovery.

Keywords: Antiphospholipid syndrome, collaborative care, gangrene, plasmapheresis, pulse therapy

Introduction

Antiphospholipid syndrome (APS) is an autoimmune disease with clinical manifestation of venous or arterial thrombosis with the persistent presence of antiphospholipid antibody (Cervera et al., 2009). Catastrophic antiphospholipid syndrome (CAPS) is the most severe form of APS also known as Asherson's syndrome. It was first discovered by Ronald Asherson in 1992. It is a life-threatening complication with occurrence less than 0.8% to 1% of patients with APS and has a mortality of about 50% (Asherson, 2005). APS is an acute and complex biological process leading to the occlusion of small vessels of various organs. Thrombotic microangiopathy multiple organ thromboses are the features seen in patients. In some patients, it causes tissue necrosis and is considered an extreme or catastrophic variant of APS (Strakhan et al., 2014). The condition presents with features of disseminated intravascular coagulopathy (DIC) or thrombotic thrombocytopenic purpura (TTP) that should be treated immediately to prevent mortality (Limper et al., 2019).

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Case details

Case presentation

The present case is a 19-year female unmarried who was bought to the emergency room in an unconscious state. She had a history of fever, chills, and rigours for three days. Her Glasgow coma scale (GCS) was eight (E2V1M5). Immediate care with ventilator support on Synchronized Intermittent Mechanical Ventilation (SIMV) with 100% fraction of inspired oxygen (FiO2), Positive End-Expiratory Pressure (PEEP) 5 cm H2O was provided. Her vitals were pulse-135 bpm, respiration – 20 bpm, blood pressure -140/80 mmHg and the temperature was 98.6°F. The patient had blackish discolouration of the great toe, 2nd and 3rd toes of the right lower limb, and in the left ankle region for three months. She had no comorbid illness and no significant past medical history.

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Relevant investigation

On admission, laboratory values were as follows- haemoglobin 9.1 gm/dl, haematocrit 28.0%, platelet count 40.0x103/μl, total white blood cells 13.7x103 /μl, neutrophils 93.3%, red blood cells 231 mg/dl, thyroid-stimulating hormone 4.470 μIu/ml, C-reactive protein 16 mg/l, NTPRO-BNP >35000 pg/ml, Troponin T 1.580 ng/ml, and 24 hour urine protein 658 mg/24 hour. Coagulation and haemostasis tests were done and prothrombin time (PT) was 14.4 sec and APTT tested with an optical method showed normal value (29.6 sec) and a test for lupus anti-coagulant showed a positive result.

Antinuclear antibody (ANA) workup showed positive APLA IgM. Anti-neutrophilic cytoplasmic antibody (ANCA) showed a 1:10 ratio, which suggested a negative result. ELISA for anti-neutrophilic cytoplasmic antibody (ANCA) titre for IgG antibody against myeloperoxidase (MPO) and proteinase 3 (PR3) and antibodies to dsDNA showed negative results. Antibodies to cardiolipin IgM titre value and ACA IgG value were positive and electroencephalogram (EEG) revealed severe diffuse encephalopathy. Electrocardiogram (ECG) was normal except for 'T' wave inversion.

2-D echo report on 3rd day of admission showed globally hypokinetic left ventricle, moderate left ventricular systolic dysfunction, severe mitral regurgitation and mild tricuspid regurgitation and diagnosed to have a congestive cardiac failure. A repeat echo on the 19th day of admission showed globally hypokinetic left ventricle, moderate to severe left ventricular systolic dysfunction, moderate to severe mitral regurgitation and right ventricular dysfunction with moderate tricuspid regurgitation. The ejection fraction was 33%.

Right vesicoureteric junction calculus and proximal ureteric calculus and minimal hydroureteronephrosis, bilateral tiny non-obstructive renal calculus and multiple para-aortic, aortocaval and retrocaval lymph nodes were seen in CT kidney ureter bladder (KUB). Plain CT brain revealed multiple ill-defined scattered areas of intra axial hypodensities involving both grey and white matter with loss of grey-white matter differentiation in cerebral hemispheres, brainstem and cerebellum suggesting multiple infarcts. Based on the above findings, the initial diagnosis was sepsis with septic shock, acute kidney injury, and suspected vasculitis and was admitted to the intensive care unit for further management.

Case management

The 19-year young women brought to the emergency room was unconscious with a Glasgow coma scale reading eight (eye-opening 2, verbal response 1, and motor response 5) and hypotension, and was managed with noradrenaline 8 mg in 42 ml in sodium chloride at 4 ml per hour. Injection Fentanyl citrate 100 mg and Ketamine 50 mg were administered intravenously during intubation and Tazillin (Piperacillin/Tazobactam) 4.5 gm in 500 ml diluted in sodium chloride intravenous bolus was administered because of septic shock. Injection sodium bicarbonate 200 ml was administered to correct metabolic acidosis. Once the patient was stabilized, she was shifted to the medical intensive care unit for further management.

In ICU, she was given Methylprednisolone a cortico-steroid 500 mg in 250 ml sodium chloride to reduce inflammation. Injection Enoxaparin sodium 0.4 ml subcutaneous was administered prophylactically to prevent deep vein thrombosis. The patient was tracheotomized given prolonged ventilation. On the 2nd day of admission, the patient had an episode of seizure for which she had received intravenous injection Levetiracetam 500 mg 12 hourly. Meropenem 1 gm intravenous 12th hourly, injection Clindamycin 600 mg intravenous 8th hourly and intravenous injection Tazillin (Piperacillin sodium/Tazobactam sodium) 2.25 gm 6th hourly was given to manage the infection. Salbutamol nebulization was given 6th hourly to treat hyperkalaemia. In the course of treatment, the patient developed anaemia for which she was transfused with packed red blood cell on the 11th and 12th day of admission. The patient was administered fresh frozen plasma on the 6th, 8th, 12th, and 15th day of admission. The patient developed heparin-induced thrombocytopenia; hence, injection Enoxaparin sodium was stopped and changed the drug to low molecular weight heparin (LMWH) Fondaperinux 5 mg

subcutaneous. The patient underwent plasmapheresis on the 6th, 7th, 9th, 13th, and 16th day of admission. High potassium levels were managed by haemodialysis on the 6th day of admission. The patient was started on Mesna 200 mg in 100 ml of sodium chloride over two hours. Simultaneously, Ondansetron 4 mg intravenous stat followed by injection Cyclophosphamide 500 mg in 500 ml sodium chloride over 6-8 hours intravenous infusion on 27th and 32nd day of admission were given. The patient was haemodynamically stable but the Glasgow coma scale was E1VTM1.

The patient had developed multidrug-resistant Klebsiella infection on endotracheal culture, which was managed by injection Tazillin (Piperacillin/Tazobactam) 4.5 gm intravenous 8th hourly in 100 ml sodium chloride. The patient developed hypertension, which was treated with tab Arkamin 100 mg QID and Amlodipine 5 mg SOS. The patient had developed gangrene of the toes, which had to be amputated to prevent further complications. In the course of the hospital stay, the patient had developed acute respiratory distress syndrome and was managed by ventilator support and frequent chest physiotherapy was given to maintain lung hygiene. Limb physiotherapy was given to prevent bed rest complications and to maintain mobility and properties of joints and muscles. The patient was weaned off from the ventilator on the 17th day of admission. With all these treatment modalities, the patient progressed to improve with GCS E4V5M6. She developed blurred vision. However, she was able to walk with support and do her daily activities of living with minimal assistance.

She was discharged with tablet Amlodipine 5 mg, tablet Clonidine 100 mg, and tablet Levetiracetam 500 mg twice daily; Elemental iron with folic acid, Prednisolone 10 mg, Calcium 500 mg with vitamin D3, and Aspirin 75 mg once a day; Alendronate Sodium 35 mg once a week, syrup Levodropropizine 2 teaspoon twice daily, and paracetamol if necessary to continue for one month. The patient was called for follow up after a month. She was also advised to consume a low salt diet and to avoid green leafy vegetables and all yellow fruits.

Interdisciplinary nursing care involved the overall well-being of the patient and family. The problems identified were congestive cardiac failure, septic shock, gangrene, and central nervous system involvement, loss of vision, acute respiratory distress syndrome, lung collapse, metabolic acidosis and acute kidney injury. Nursing care was delivered based on the problems identified as shown in Table 1.

The catastrophic antiphospholipid antibody is a syndrome of multiple vascular occlusions associated with high titres of antiphospholipid antibodies (APLA). Presentation of catastrophic antiphospholipid antibody syndrome is often complex as it involves multiple organs over a short period typically a few days to weeks (Oliveira, Correia, & Oliveira, 2020). The most characteristic involvement in catastrophic antiphospholipid antibody syndrome is of renal, pulmonary, cerebral and gastrointestinal vessels. The central nervous system is involved and is manifested by the presence of cerebral infarctions. Micro thrombi and micro infractions are observed in catastrophic antiphospholipid antibody syndrome which results in thrombotic events in microcirculation leading to gangrenes and amputation (Asherson et al., 2001). The present case also showed similar findings involving multiple infarcts in the brain, gangrene in the foot requiring amputation of the toes of the left foot.

The present case had high potassium and urea, which was treated with haemodialysis. The patient had undergone five cycles of plasmapheresis because of high titres for ANA. The dexamethasone- cyclophosphamide pulse (DCP) therapy regimen was started as there was no improvement with plasma-pheresis. The treatment involved intravenous administration of Mesna before starting DCP to prevent bladder bleeding and anti-emetic to prevent nausea and vomiting which is a complication of DCP. Post administration of DCP, the same regimen was followed and the patient had shown significant improvement with a decrease in the symptoms. In the course of the treatment, the patient had developed left lung collapse, which was recovered with ventilator management and chest physiotherapy. There was a significant improvement in the level of consciousness and the general condition of the patient with the ability to carry out the activities with minimal assistance. As challenging as the case may be, we have seen a significant improvement in the condition of the

patient clinically. Presently the patient was conscious, oriented and able to move in bed. The patient was discharged from the hospital while she was able to carry out her activities with minimal assistance. Comprehensive nursing care involved preventing bed rest complications, hospital-associated infections, and bowel and bladder dysfunction by providing the patient with oral care, tracheostomy care, and psychological support for both patient and family members. An interdisciplinary approach is crucial in improving the patient's condition.

Table 1: *The systems affected and the care provided*

System affected	Cardiovascular system	Central nervous system	Respiratory system	Renal system
Problems identified	· Congestive cardiac failure (CCF) · Gangrene · T-wave inversion · Septic shock	· Altered GCS · Encephalopathy · Motor and sensory impairment · Vision loss	· Infection · Acute respiratory distress syndrome · Lung collapse	· Metabolic acidosis · Renal calculi · Acute kidney injury
Focus of care	✓ Decreased intravascular volume: Administration of IV fluids and monitor fluid status ✓ Decreased preload and decreased afterload: Monitor fluid and electrolyte status through laboratory investigations, administer oxygen, maintain intake and output chart. ✓ Improving gas exchange and oxygenation ✓ Reduce anxiety ✓ Balancing rest and activity ✓ Monitoring for cardiac arrhythmias	✓ Monitoring ICP ✓ Assessing GCS ✓ Prevention of pressure sores ✓ Providing other means of communication ✓ Maintaining joint mobility by passive range of motion exercise	✓ Monitoring respiratory state ✓ Humidification ✓ Ventilator support and care ✓ Positioning ✓ Nebulisation ✓ Chest physiotherapy	✓ Maintaining intake and output ✓ Correction of electrolyte imbalance ✓ Monitoring arterial blood gas analysis ✓ Care during haemodialysis
Outcome	* CCF resolved * Gangrene treated with amputation * No new arrhythmias * Recovered from septic shock	* Improved GCS score to 15 * Treated encephalopathy * Improved motor and sensory impairment * Blurred vision	* Free from infection * Recovered from ARDS * Good air entry to bilateral lungs	* Corrected metabolic acidosis * Renal calculi still present * Reverted AKI to normal function

Conclusion

In conclusion, catastrophic antiphospholipid antibody syndrome in the present case has involved the brain, kidney, heart and lungs. Treatment strategies with the support of mechanical ventilator, haemodialysis, plasmapheresis, and immunoglobulin high standard collaborative care has given a better result though the literature shows poor prognosis.

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References

Asherson R. A. (2005). The catastrophic antiphospholipid (Asherson's) syndrome in 2004--a review. Autoimmunity reviews, 4(1), 48–54. https://doi.org/10.1016/j.autrev.2004.03.007

Asherson, R. A., Cervera, R., Piette, J. C., Shoenfeld, Y., et al. (2001). Catastrophic antiphospholipid syndrome: clues to the pathogenesis from a series of 80 patients. Medicine, 80(6), 355–377. https://doi.org/10.1097/00005792-200111000-00002

Cervera, R., Bucciarelli, S., Plasín, M. A., et al. & Catastrophic Antiphospholipid Syndrome (CAPS) Registry Project Group (European Forum on Antiphospholipid Antibodies) (2009). Catastrophic antiphospholipid syndrome (CAPS): Descriptive analysis of a series of 280 patients from the "CAPS Registry". Journal of autoimmunity, 32 (3-4), 240–245. https://doi.org/10.1016/j.jaut.2009.02.008

Limper, M., de Leeuw, K., Lely, A. T., et al. (2019). Diagnosing and treating antiphospholipid syndrome: A consensus paper. The Netherlands journal of medicine, 77(3), 98–108.

Oliveira, D. C., Correia, A., & Oliveira, C. (2020). The Issue of the Antiphospholipid Antibody Syndrome. Journal of clinical medicine research, 12(5), 286–292. https://doi.org/10.14740/jocmr4154

Strakhan, M., Hurtado-Sbordoni, M., Galeas, N., Bakirhan, K., Alexis, K., & Elrafei, T. (2014). 36-year-old female with catastrophic antiphospholipid syndrome treated with eculizumab: A case report and review of literature. Case reports in haematology, 2014, 704371. https://doi.org/10.1155/2014/704371