

Dramatic response to plasma exchange in systemic lupus erythematosus with acute complications: Report of two cases

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Abstract

Acute exacerbations and complications are common in patients of systemic lupus erythematosus (SLE) despite of adequate long-term therapy with immunosuppressive drugs. So other options like therapeutic plasma exchange (TPE) can be considered as part of management. Here, we share our experience of two patients of SLE with acute complications who were successfully managed with TPE.

Keywords: Diffuse alveolar hemorrhage, systemic lupus nephritis, therapeutic plasma exchange

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Introduction

Systemic lupus erythematosus (SLE) is an acquired immunological disorder in which tissue damage is mediated by autoimmune antibodies and immune complexes. Patients with SLE usually have chronic debilitating course requiring long-term immunosuppressive therapy even then acute exacerbations and complications are common. Acute flares/complications are usually treated by pulse methylprednisolone therapy. However, some patients do not respond to pulse steroid therapy and may be candidates for other treatment options like therapeutic plasma exchange (TPE). Although, TPE in lupus nephritis is classified as category IV indication, it is considered as category II indication when SLE is associated with diffuse alveolar hemorrhage.^[1]

Case Reports

Case 1

A 28-year-old female presented with history of

shortness of breath, cough with blood stained sputum and oliguria for 1 day. There was history of pedal edema, multiple joint pains associated with swelling for 10 days, with history of oral ulcers off and on and polymenorrhagia for 6-8 months. There was also history of untreated mild to moderate hypertension for 2 years. She was admitted in intensive care unit (ICU) for ventilator support. On examination, Jugular Venous Pressure (JVP) was raised with severe pallor and pedal edema. Auscultation of chest revealed fine crackles all over the chest. Abdominal examination showed hepatomegaly. X-ray and computed tomography (CT) scan of chest showed diffuse alveolar opacities in bilateral upper and middle lobes of lungs. Initial urea and creatinine values were 89 mg/dl and 2.2 mg/dl respectively. Serology revealed positive ANA and anti double strand DNA antibodies. Patient was diagnosed as SLE with acute flare up in the form of diffuse alveolar hemorrhage and renal failure. Patient was intubated and put on a ventilator, started with pulse methylprednisolone therapy and injection cyclophosphamide. Three units of packed red cells were transfused for severe anemia (Hb-4.7 g/dl). As there was no improvement after 48 h, the treating physician planned plasma exchange for the patient. The patient was assessed for the procedure by transfusion medicine team and the patient was shifted to department

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of transfusion medicine on ventilatory support. One plasma volume of patient was exchanged with normal saline and 4% human serum albumin as replacement fluid. No procedural complications were observed and he was shifted back to ICU. After 24 h of the procedure, patient was successfully weaned off the ventilator and extubated with no fresh episodes of hemoptysis. However, urine output was still low with hematuria. A total of four plasma exchange cycles were done for the patient on alternate days and one plasma volume was exchanged each time. After the 2nd procedure, there was an improvement in urine output without any need for renal replacement therapy. After the 3rd procedure, urine output was normal without hematuria, with no fresh episodes of hemoptysis and a repeat X-ray showed clearing of hemorrhagic opacities from lungs. Patient was discharged after the 4th procedure, though azotemia persisted. Her total duration of hospital stay was 14 days with ICU stay of 10 days.

Case 2

A 24-year-old male, a known case of SLE (for 13 years) with End Stage Renal Disease (class IV lupus nephritis) presented with shortness of breath, chest pain, and cough for 7 days. Patient had two episodes of diffuse alveolar hemorrhage in the past, managed with pulse steroids. The patient also had pulmonary tuberculosis and was on Anti-tubercular treatment for last 6 months. He was also Hepatitis B surface antigen (HBsAg) positive. He had received five pulses of cyclophosphamide prior to this admission. On examination, patient was tachypnoeic, endotracheal bleeding was present. There was right hypochondriac tenderness and hepatomegaly. He was admitted to ICU for ventilator support and was started on pulse methylprednisolone. Chest X-ray and CT scan showed bilateral middle and lower alveolar opacities. The possibility of acute exacerbation of SLE in the form of diffuse alveolar hemorrhage was considered. As there was no improvement on pulse steroids, plasma exchange was planned. Patient was assessed by transfusion medicine team and taken up for the plasma exchange on day 2 of ICU admission. Six cycles of plasma exchange were administered on alternate days by exchanging one plasma volume, using 4% human Serum Albumin as replacement fluid. After the 1st procedure there was clearing of the endotracheal tube bleeding and after the 2nd procedure patient could be successfully weaned off the ventilator and was shifted to the ward. Patient was on dialysis simultaneously and his renal function tests improved after the 6th procedure. His total duration of hospital stay was 17 days including 4 days in the ICU.

Discussion

Alveolar hemorrhage is uncommon in SLE, occurring in less than 2% of the patients. It accounts for 1.5-3.7% of hospital admissions due to SLE. However, it may be a life-threatening complication with high mortality rate of 53-86%.^[2] In our 1st case, patient presented with diffuse alveolar hemorrhage as the initial symptom of the disease and in the 2nd case patient was already a diagnosed case of SLE having recurrent alveolar hemorrhage. Both cases needed immediate ventilator support and were given rescue steroid therapy without any improvement. Hence, plasma exchange was planned as desperate life saving measure. Plasma Exchange has been found to be effective in numerous case reports.^[3,4] Rationale suggested for effectiveness of plasma exchange is that it clears immune complexes from the circulation which mediate cascade of events leading to these complications in patients of SLE.^[5,6]

In previous reports, plasmapheresis was instituted if the patients had an inadequate clinical response to high-dose corticosteroid and cyclophosphamide therapy as was done in our cases. Problems with plasmapheresis are the theoretical possibility of increased autoantibody synthesis as a "rebound phenomenon" and infection of the intravenous line. The auto antibody production can be decreased by giving a cytotoxic agent before plasmapheresis.^[7] As our patients were started with immunosuppressive drugs prior to start of plasma exchange, response in the form of resolution of alveolar hemorrhage was good in the first two cases and patient could be weaned off the ventilatory support within 24-72 h.

There is still controversy about the efficacy of plasma exchange in the treatment of severe lupus nephritis as seen in the 2nd case, which needed dialysis in addition to plasma exchange. Some studies have reported no beneficial effect with plasma exchange, while others claim its usefulness when used in combination ("synchronization") with immunosuppressive drugs, particularly intravenous pulse doses of cyclophosphamide. In our case, immunosuppressive drugs were continued with plasma exchange along with dialysis and renal functions improved after the 6th cycle. According to another case report, the addition of plasma exchange to intravenous pulse cyclophosphamide might be effective in patients with severe lupus nephritis unresponsive to immunosuppressive drugs alone.^[8] However, Lewis, et al. in a controlled trial involving 86 patients with severe lupus nephritis, reported that plasmapheresis plus a standard regimen of prednisone and oral cyclophosphamide did not improve the clinical outcome of these patients as compared with the standard regimen alone.^[9]

To conclude, clinically significant improvement in the patients described above after plasma exchange suggest that it can be an important component of treatment in patients of SLE with acute life threatening complications in addition to conventional high dose steroid and cytotoxic drug therapy.

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