中文題目：免疫球蛋白在紅斑性狼瘡合併敗血症及低CD4淋巴球治療-個案報告

英文題目： IVIG Treatment in SLE with sepsis and low CD4 lymphocyte counts – A Case Report

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Introduction:

Systemic lupus erythematosus is a disease difficult to diagnose and can be diagnosed by criteria only. We would like to present a young man with history of proteinuria without regular follow-up and suffered from generalized edema for several months.

Case report:

A 19-year-old man complained of general weakness, dyspnea, pale looking, and dizziness while standing in recent 1 week; and spiking fever for 2 days. He was brought to our ER on 2011/7/18 by his friend because of near fainting. At our ER, his consciousness was clear, but in acute respiratory distress appearance, pale looking, breathing sounds revealed bilateral crackles. Laboratory examination data revaeled severe anemia and thrombocytopenia, leukocytosis, elevated CRP(13.6 mg/dl) and PCT level(45.66 ng/uL), renal function impairment, and severe hypoalbuminemia. Chest film showed bilateral pleural effusion and he was admitted to MICU.

After admission, blood transfusion including PRBC and PLT were done for severe anemia (HgB:1.6mg/dl) and thrombocytopenia (5000/uL). 24 hours urine protein: 11.2g and serum albumin was only 1.7 mg/dl. Infection specialist suggested empirical use of PCN-G to cover Leptospirosis, Minocin to cover Q-fever and scrub typhus infection, and Rocephin to cover Grams negative bacteria infection. Solucortef 100mg q8h was prescribed for sepsis. Both direct and indirect Coombs' test showed positive. Blood smear showed spherocytosis without fragmented RBC. Serum analysis reported ANA(+)160x speckled pattern, anti-cardilipin Ab IgM(+), low C3/C4, low IgG (531 mg/dl), but negative of other ENAs, anti-B2-GP-I Ab, ANCA, lupus anticoagulant, and RPR. Methylprednisolone 80mg daily was prescribed and we escalated antibiotics from Rocephin to Tienam for severe sepsis.

Bilateral pig-tail chest drainage tubes were inserted for empyema drainage under platelets transfusion on the 3rd hospital day. Both blood cultures and pleural effusion culture revealed *Klebseilla pneumoniae* later. Patient's clinical status, hemolytic anemia, thrombocytopenia, empyema, serum creatinine, and sepsis improved after antibiotics and systemic corticosteroid use. Steroid was tapered gradually (7/20~7/23 MTP 80mg dailly; 7/24~7/28 MTP 60mg daily; 7/29~7/31 MTP 40mg daily). Virus titers including HIV and HTLV-1 were checked and were all negative. Otherwise, Eltroxin was given due to hypothyroidism (T4:1.612 TSH:10.312) and bone marrow biopsy revealed no evidence of leukemia or macrophage activation syndrome. He was

transferred to AIR ward on 7/28.

Tienam 1g q8h for 14 days (2011/7/21-8/2) and checked PCT was normal. Platelet count remained around 100000 to 110000 uL from 7/75~7/28. On 2011/8/1, we tapered down methylprednisolone to oral prednisolone 15 mg BID and platelet count decreased from 82000 ul to 59000 ul (8/4). Thus, we adjusted corticosteroid to methylprednisolone 80 mg QD and bilateral pigtail drainage were removed on8/4. Albumin supplement and diuretics were used for bilateral legs edema. Immunoglobulin was checked which revealed reduced IgG(531->288 mg/dl) with IgM and IgA in normal range. On 8/6, Baktar 2# qd prophylaxis was prescribed due to low CD4 count (72 cells/uL).

On 8/7, spiking fever with chills developed with normal PCT. Yet, another sepsis was suspected, so Tienam was prescribed empirically after blood & urine cultures. Pyuria was found and review of PB smear revealed positive of schistocytes (1-2 schistocytes/HPF); high D-dimer & decreased of fibrinoggen, so DIC was diagnosed. Due to clinically suspicion of another sepsis and immunecompromise, IVIG was given on 8/7 and follow up IgG elevated to 687 mg/dl together with the improvement of sepsis and platelet count. Later, multiple drugs resistant Acinectobacter baumanni was cultured. Gallium scan revealed active inflammation in both kidneys and suspicious focal inflammation in RLQ abdomen (cecum or A-colon). Stool routine was checked which revealed no pus cell or occult blood; PE revealed no tenderness or rebounding pain. Abdominal sonography revealed bilateral enlarged kidney with poorly differential echogeneitiy in between cortex and medulla (chronic kidney disease) together with fatty liver and type V membranous nephritis was reported via renal biopsy. Thrombocytopenia improved on 8/19 (plt:62000) and serum creatinine was within normal limit after Tienam and amikin used for 14 days (8/7-8/21). Under relative stable condition, finally, he was discharged on 8/22.

Conclusion:

The final diagnoses were listed below:

1. Sepsis, first time of sepsis was pneumonia with *Klebsiella pneumoniae* empyema and bacteremia (Jul.21, 2011~Aug.2, 2011); the second time was urinary tract infection with *Acinectobacter baumanni* bacteremia , complicated with disseminated intravascular coagulopathy (Aug.07, 2011~Aug.21,2011).
2. Systemic lupus erythematosus [ANA(+): 160X speckled pattern, autoimmune hemolytic anemia, thrombocytopenia, anticardiolipin antibody poitive, and lupus nephritis with nephrotic syndrome)
3. Immunocompromised with low CD4 count (72 cells/uL) and low IgG s/p self-paid IVIG with Bactar prophylasis (Aug.7, 2011)
4. Autoimmune hemolytic anemia with idiopathic thrombocytopenic purpura (suspected Evan's syndrome), s/p bone marrow biopsy; R/I systemic lupus erythematosus related.
5. Hypothyroidism, under Eltroxin treatment

However, since TTP & HUS were not likely. AIHA combined with ITP (Evan's syndrome) were considered and with suspicion of autoimmune disease related. But if we think the thrombocytopenia as one presentation of SLE, Evan's syndrome should not be considered because it was not ITP. Immunocompromised status was noted on this patient and after IVIG treatment, clinical condition became stable. Through this case, we got a conclusion that IVIG treatment may be helpful in patients with SLE and low CD4 count during active infection.