Immunoadsorption for Children systemic lupus

Ren Qi¹ YU ShengYou²

1. Tongji Medical College of Huazhong University of Science and Technology, HuBei, WuHan, 430030, China
2. Central Laboratory1022, Guangzhou Medical University, Guangzhou, Guangdong, 510180, China

E-mail: shengyouyu@163.com

Immunoadsorption (IAS) is used as a rescue therapy in SLE patients who are refractory to conventional therapies. IAS is superior to other related therapies, such as plasma exchange. In addition, prolonged IAS appeared beneficial without increasing the risk for side effects. This aims at the rapid and extensive removal of pathogenic immunocomplexes (ICs) and (auto-)antibodies (Abs). IAS can decrease the titers of ANA and ds-DNA antibody. We found that it can decrease the activity of SLE and is of higher safety. IAS offers an alternative therapeutic strategy in severe, active refractory SLE not only in the short-term. IAS was safe and effective in our study.


Key word: Immunoadsorption; systemic lupus; immune complexes; Children

Introduction

SLE is characterized by pathogenic autoantibodies, which can be removed by extracorporeal procedures. So, pathogenic autoantibodies are a hallmark of SLE. They can bind to cells and tissues, inducing complement activation and severe inflammation in the affected organ [1]. Anti-double-stranded DNA antibodies (anti-dsDNA) are associated with lupus nephritis [1–9]. Inhibiting pathogenic autoantibodies can prevent their pathogenetic consequences. In fact, immunosuppression aims at interfering with autoimmune and IC formation. In contrast to plasma exchange, IAS allows for the specific and nearly complete clearance of circulating Ig and IC, while neither removing other plasma proteins nor necessitating substitution with fresh frozen plasma, albumin or immunoglobulins[10-12]. In severe SLE with major organ involvement, the therapy goal is to stabilize disease activity at low levels. IAS appeared relatively safe, with infectious adverse events in the range of a matched group of similarly active SLE patients [13]. IAS is feasible in severe SLE, and in complicated situations with limited therapeutic options, such as, in particular, in pregnancy[14], in active tuberculosis under triple therapy[10], or in patients with antiphospholipid syndrome (APS)[15].

Case Report

A ten-year-old women children admitted with a chief complaint of "2 week fever, 5 days edema". Physical examination: T:38.7°C, P 120 per min, R 25 per min, BP:134/91mmHg, face edema, erythema on zygomatic region, pharyngeal hyperemia, no abnormal in heart and lung, abdomen is flat, liver is 3cm under the right ribs and spleen were impalpable, both lower limb edema. Bloody examination: hemoglobin is declined (54g/L), platelet is declined (56×10⁹/L). Urine routine examination: RBC (++++), urine protein> 4.10 g / L, ESR 83 mm / h. Complement is declined. Cardiac Function, liver function and kidney function are abnormal. Antinuclear antibody is increased. Anti-dsDNA antibody is positive. Lupus cell is found in peripheral blood. Admitted to hospital 4 days later,
the children was diagnosed as "SLE and lupus nephritis (LN)". Given methylprednisolone continuous stosstherapy for three courses, and then given combined therapy with hormone and immunosuppressants for 30 days, the patient child’s disease condition was not improved obviously, but was progressive severe. Joint pain and gross hematuria were occurred on patient child.

We tried to carry out IAS to treat severe Children Systemic Lupus Erythematosus. We used DNA 280 Immunity Adsorption hemoperfusion device to operate whole blood absorption. We observed the effect of cleaning up varietal own antibodies, at the same time, observed the clinical symptoms and situation of proteinuria. We operated whole blood absorption a total of two times at interval of 4 days. The operation was gone smoothly and no adverse reaction was found. After IAS treatment, the clinical symptoms was improved obviously. The body temperature and blood pressure became normal. The skin erythema in face and zygomatic region was improved obviously. Joint pain and gross hematuria disappeared. Hemoglobin and platelets are increased to normal. Urinary protein and urine erythrocyte is reduced obviously. Heart function, liver function and kidney function are improved obviously. Complement is increased. The titer of antinuclear antibodies (ANA) is decreased after the first treatment. The ANA was decreased obviously and Anti-dsDNA antibody became negative after the second treatment. With 30 days admission, the patient discharged with improvement so that shorten the length of hospitalization and reduced the family burden.

IAS, which we used in this study, received good efficacy and deserves wide clinical application.

**Discussion**

Extracorporeal therapies are a rescue strategy in critically ill SLE patients when conventional strategies have failed or are contraindicated [16]. IAS is still experimental, those patients finally undergoing IAS are characterized by active and progressive SLE resistant to conventional treatment. Thus, interpreting therapeutic effects in these patients on a background of previous immunosuppressive therapy is difficult. For the IAS procedure, blood is taken from a peripheral vein and plasma and corpuscular elements are separated by centrifugation. Then, the plasma slowly flows over adsorption columns and Ig and IC are bound via specific ligands. In most cases, two columns are assigned for each patient. During IAS, one column at a time is in use while the other one is cleaned of bound Ig; between IAS sessions, the columns are stored under sterile conditions. IAS has following advantages: ① Specific combination of antigen and antibody have high selectivity and specific adsorption in a variety of autoantibodies; ② plasma components did not lost and strength of treatment is adjustable to disease situation; ③ No replacement fluid so that eliminate the risk of disease infection; ④ Operation is simple, and treatment is efficient. IAS is one new technology for SLE treatment, IAS is characterized by good efficacy, no obvious toxic and adverse reaction. It is suitable for wide clinical application.

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**Corresponding Author:**

Dr. YU SY
Guangzhou Medical University
Guangzhou, Guangdong, 510180, China
E-mail: shengyoyyu@163.com
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