INTISARI

Autoimmune Hemolytic Anemia (AIHA) merupakan kondisi terbentuknya antibodi terhadap sel darah merah sehingga menyebabkan proses hemolisis. Pada pasien talasemia beta mayor yang mengalami transfusi berkala memiliki risiko terjadinya AIHA sebesar 5 dari 100 pasien talasemia, hal ini lebih besar dibandingkan populasi umum.

Anak perempuan usia 15 tahun dengan diagnosis AIHA berulang dan talasemia beta mayor. Pasien telah mendapatkan terapi steroid sebelumnya, tetapi tidak remisi. Pada laporan kasus ini, pasien ditatalaksana dengan pemberian siklosporin dosis rendah dan steroid, serta pemberian PRC *leukodepleted*. Remisi AIHA tercapai hingga satu tahun pemantauan.

Tatalaksana AIHA berulang dapat menggunakan siklosporin dosis rendah dan steroid, disertai tatalaksana non farmakologis berupa transfusi dengan PRC *leukodepleted* dan pembentukan kelompok donor transfusi. Meskipun pada penelitian ini kelompok donor yang terbentuk tidak memenuhi kebutuhan darah pasien.

Kata kunci : Talasemia, AIHA berulang, siklosporin, PRC *leukodepleted*

ABSTRACT

Introduction and importance: Autoimmune hemolytic anemia (AIHA) is characterized by the production of antibodies against the red blood cells (RBCs) leading to increased hemolysis. Incidence of AIHA in patients with beta-thalassemia major is 5 of 100 thalassemia patients.

Case presentation: A 15-year-old female patient with refractory AIHA and thalassemia was unresponsive to steroid therapy. In this report, she was treated with cyclosporine and prednisone in addition to leucodepleted packed red cell (PRC). She was successfully got remission during one-year monitoring.

Clinical discussion: Cyclosporine is an immunosuppressive agent that interferes with T-cells activation by inhibiting transcription of cytokines, such as interleukin 2 and interferon-ɣ. Cyclosporine and steroid have synergic effects to prevent antibody production. Nephrotoxicity is one of the most concerning effect in cyclosporine usage, but it rarely develops using doses lower than 5 mg/kgBW/day. In limited resources blood bank with ABO and Rh crossmatch only, leucodepleted PRC transfusion could be an effective way to prevent antibody formation to minor blood group.

Conclusion: Cyclosporine and steroid could be considered for management in refractory AIHA with thalassemia patients. Non-pharmacological therapy such as leucodepleted PRC transfusion and limited donor transfusion could be considered.

Keywords: thalassemia; refractory AIHA; cyclosporine; leucodepleted PRC; case report