

Purification treatment with Cytosorb® as a bridge to emapalumab in a severe case of macrophage activation syndrome secondary to systemic lupus erythematosus

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Introduction

We report the case of a young girl with a past medical history of vertically transmitted C hepatitis, treated with ledipasvir/sofosbuvir at the age of 12, obtaining a persistent negativization of viraemia.

Methods

At 14 years of age she was diagnosed with systemic lupus erythematosus with class IV-V lupus nephritis determining anuric AKI that required hemodialysis for 3 weeks. She received hydroxychloroquine, high dose steroids, i.v. cyclophosphamide, plasma-exchange and showed good recovery of renal function. She developed thrombotic microangiopathy and was therefore started on eculizumab, showing a good clinical response. Three months after the disease onset, while she was on maintenance therapy with prednisone, hydroxychloroquine and eculizumab, she was re-hospitalized for severe fluid overload and diagnosed with macrophage activation syndrome. She showed very poor response to standard treatment with high dose steroids and anakinra; as a rescue therapy emapalumab was therefore requested, but was not immediately available.

Results

Considering the very severe clinical presentation (anasarca in pseudo pseudo Meigs syndrome secondary to SLE, lymphopenia, hypofibrinogenemia, hyperferritinemia) with still impaired renal function, a double-lumen CVC was placed and an extracorporeal blood purification treatment with Cytosorb was carried on for 24 hours with a stand-alone application on a Prismax RRT machine; hemodynamics remained stable and no side effects were observed. The patient was then started on emapalumab, showing a good clinical response with complete resolution of macrophage activation syndrome within 6 weeks.

Conclusion

After 8 months period of follow up she is showing no disease activity with normal ferritine, negative anti DS DNA antibodies and normal glomerular filtration rate (110 ml/min/1.73mq).

