AB0929

ACUTE RHEUMATIC FEVER: IS PROLONGED CORTICOSTEROID TREATMENT ASSOCIATED TO A BETTER CARDIAC OUTCOME?

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Background: Acute rheumatic fever (ARF) is a nonsuppurative sequela that can occur two to four weeks after group A beta-hemolytic Streptococcus pharyngitis. Despite its decline in incidence in modern countries, ARF still represents worldwide a serious healthcare concern. The most common manifestations are arthritis and carditis, followed by chorea, erythema marginatum, and subcutaneous nodules. The diagnosis of ARF is clinical and requires satisfaction of revised Jones criteria as well as evidence of a recent streptococcal infection.

Carditis and chorea both need systemic corticosteroid treatment (prednisone, 1 mg/kg) but with different treatment courses: 2-3 weeks prednisone treatment course in carditis is generally undertaken compared to a longer prednisone treatment period in chorea (at least 2 months).

Objectives: The aim of this study was to investigate if the longer corticosteroid treatment used for patients with chorea and carditis is related to a better regression of cardiac damage compared to the shorter course of corticosteroid treatment in patients with only carditis.

Methods: Data regard 14 patients were retrospectively revised (8 males, 6 females): 7 ARF patients with both cardiac and neurological involvement treated with systemic prednisone for 2 months; 7 ARF patients with only cardiac involvement treated with systemic prednisone for 2 weeks. The regression of cardiac damage in the two different groups was evaluated in relation to the different duration of corticosteroid treatment comparing ultrasound echocardiography (cardiac-US) performed at ARF onset and during a 2-3 year follow-up. The trend of the grade of aortic and mitral valves insufficiencies was chosen as indicator of the ARF cardiac damage evolution.

Patients with a similar grade of cardiac damage at disease onset were identified; then pairs were created according to the presence of chorea (so one patient was affected only by carditis and the other one of the same pair, according to the similar cardiac damage, had both carditis and chorea). Cardiac-US damage regression of the 7 pairs was assessed in order to evaluate if the grade of regression was higher in the patients with only carditis compared with the ones with both carditis and chorea.

Results: in 3 pairs a better regression of the cardiac damage was evident in the patients with only carditis; in other 3 a better regression was noticed in the patients with both carditis and chorea; finally no difference was detected in the last pair. No significant cardiac-US difference according to patient pairs appeared in relation to the steroid time treatment course (2-3 weeks vs 2-3 months); however in 85% of the cases (12 patients) steroid therapy did achieve regression on ARF cardiac damage.

Conclusion: This retrospective study does not reveal any significant difference between steroid duration therapies in ARF complicated by carditis; limits of the study are the size sample as well as the intra/inter observer variation during cardiac-US (all exams were achieved in a high level experience paediatric cardiac service); this study underlines the established importance of the use of corticosteroid in ARF carditis.

Disclosure of Interests: None declared

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AB0930

EFFICACY OF TOCILIZUMAB IN TREATMENT OF SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS IN PATIENTS WITH MACROPHAGE ACTIVATION SYNDROME

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Background: Macrophage activation syndrome (MAS) is a rare and severe complication of juvenile idiopathic arthritis (JIA) that most typically develops in patients with its systemic form (sJIA). Tocilizumab proved to be efficacious for treatment of both systemic JIA and polyarthritis. Nevertheless, the question regarding the effect of MAS on efficacy of TOC therapy in patients with sJIA is still to be solved.

Objectives: To evaluate the efficacy of tocilizumab therapy in patients having sJIA and MAS versus the patients without MAS.

Methods: Throughout the period between 2009 and 2018, 251 children with the acute phase of sJIA started to be treated with TOC at the Rheumatology Department of the National Medical Research Center of Children's Health (Moscow, Russia). Of those, 69 patients had active MAS at initiation of TOC therapy. Patients were diagnosed with MAS according to the preliminary diagnostic guidelines for MAS complicating sJIA [1]. Treatment efficacy was evaluated according to the dynamics of clinical and laboratory signs using the ACRPed criteria. The Wallace criteria were used to evaluate whether or not remission had been achieved.

Results: At initiation of TOC therapy, patients with and without MAS were comparable in terms of their sex and age characteristics, as well as the overall severity of arthritis (the percentage of women, 56.6% (intergroup $p=0.396$); the median age, 7.83 (4.39-11.3) years (intergroup $p=0.640$); and JADAS-71, 17.4 (13.7-22.8) (intergroup $p=0.055$)). Nevertheless, patients differed in terms of intensity of certain clinical manifestations and prior history of the disease. The group with MAS was characterized by such parameters as shorter disease duration, shorter duration morning stiffness, and smaller number of affected joints but higher activity and severity score (Patient/Parent and Physician VAS score).

After treatment initiation, TOC was discontinued after treatment for 4-12 weeks only in 2 out of 69 (2.9%) patients because of persistent MAS or MAS flare. The patients successfully achieved remission after therapy switching. No intergroup differences were observed for the rate of treatment discontinuation within the first year of therapy: the drug was discontinued in 5 (7.2%) patients in the MAS+sJIA group (2 MAS flares + 3 cases of poor efficacy) and in 25 (13.7%) patients in the sJIA group (4 SAEs + 21 cases of poor efficacy).

TOC efficacy in sJIA depending on MAS presence

Figure 1

One-year treatment with TOC proved that this drug is highly efficacious to treat sJIA regardless of whether or not patients had MAS at treatment initiation (Figure).

No deaths were reported throughout the entire treatment period. We attributed this to early diagnosis and high efficacy of treatment of JIA patients.

Conclusion: Tocilizumab therapy in patients with sJIA in the presence of MAS is as efficacious as in patients without MAS and does not aggravate the course of MAS.

REFERENCE

- [1] Ravelli A, Magni-Manzoni S, Pistorio A, Besana C, Foti T, Ruperto N, Viola S, Martini A. Preliminary diagnostic guidelines for macrophage activation syndrome complicating systemic juvenile idiopathic arthritis. *J Pediatr*. 2005;146(5):598-604.

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