AB0749

LABORATORY MARKERS OF INFLAMMATORY ACTIVITY IN PATIENTS WITH MONOGENIC AND POLYGENIC AUTOINFLAMMATORY DISEASES

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Background: Auto-inflammatory diseases (AIDs) – a group of diseases of monogenic and polygenic nature, manifested by recurrent attacks of systemic inflammation in the form of fever, accompanied by multi-system symptoms, which are based on the pathology of natural immunity in the absence of autoantibodies and markers of infection.

Objectives: to evaluate the serum levels of markers of systemic inflammation in patients (pts) with monogenic AIDs (FMF, CAPS, TRAPS) and polygenic AID-systemic Juvenile Arthritis (sJA).

Methods: In 85 pts (68 children and 17 adults) with monogenic AIDs and sJA, the concentrations of C-reactive protein (CRP), serum amyloid A (SAA) and ferritin in the blood serum were detected. SRP and SAA were determined by the nephelometric method using Siemens reagents on a BN Prospect Siemens analyzer. Normal CRP values are 0-5 mg/L, normal SAA values are 0-5 mg/L. Ferritin was determined by ELISA using kits manufactured by Orgentec Diagnostica GmbH (normal values of 7-142 µg/L). During statistical processing of the material, the median (Me), lower and upper quartiles (LQ and UQ), minimum and maximum values were evaluated, and the reliability of differences between the groups was evaluated using the Mann-Whitney U criterion. The correlation between the values of CRP and SAA, CRP and ferritin was estimated by the Pearson criterion.

Results: The study included 30 with systemic JIA, FMF-17 pts, CAPS 25- pts and TRAPS- 17 pts. The results of the detection of CRP, SAA, and ferritin are presented in Table 1.

Table 1. CRP, SAA, and ferritin values in patients with sJA, FMF, CAPS and TRAPS.

Value	sJA	FMF	CAPS	TRAPS
CRP (N=0-5 mg/L)				
Me (LQ - UQ)	6,95(0,5 - 51,7)	30,8(9,41-47,4)	4,61(0,36-20,4)	5,41(2,77-14,7)
Min	0,15	0,48	0,15	0,15
Max	229,0	109,0	80,3	90,1
SAA(N=0-5 mg/L)				
Me (LQ - UQ)	19,05(3,05-257,0)	85,2(38,4-276,0)	7,83(2,25-122,0)	10,6(5,7-72,7)
Min	0,88	1,58	0,88	0,88
Max	962,0	923,0	346,0	516,0
Ferritin (N=7-142 µg/L)				
Me (LQ - UQ)	61,09(22,03-385,76)	57,22(21,36-133,51)	20,49(8,13-28,88)	16,52(5,42-67,27)
Min	0,73	3,36	0,98	2,42
Max	1500,0	174,22	183,57	930,1

Serum concentrations of CRP that exceeded the reference values were observed in 15(50%) pts with sJA, 11(65%) pts with FMF, 17(65%) pts with CAPS, and 5 (29%) pts with TRAPS. An increase in SAA was observed in 23(77%) pts with sJA, 12(71%) pts with FMF, 17(68%) pts with CAPS, and 8 (47%) pts with TRAPS. An increase in ferritin was observed in 9(30%) pts with sJA, 1(6%) pts with FMF, 1(4%) pts with CAPS, and 1 (6%) pts with TRAPS.

A statistically significant difference in the Mann-Whitney U test for SAA concentration was found only between the CAPS and FMF groups ($P=0.03$), and in the CRP concentration also only between the CAPS and FMF groups ($P=0.009$). The strongest statistically significant correlation according to the Pearson criterion in the whole study group was noted for CRP and SAA: $r=0.74$ ($p=0.0000$), which corresponds to the average correlation strength, a slightly weaker correlation was noted for the pair of CRP/ferritin values: $r=0.53$ ($p=0.0000$), which also corresponds to the average correlation strength. For the SAA/ferritin pair as a whole, the lowest correlation was observed in the studied group: $r=0.36$ ($p=0.0008$). The maximum correlation of studied parameters was observed in the group of patients with TRAPS: for the CRP/SAA pair, it was $r=0.9$ ($p=0.00003$), for the SAA/ferritin pair it was $r=0.7$ ($p=0.0072$), and for the CRP/ferritin pair, $r=0.59$ ($p=0.0345$). For FMF, there was a strong correlation in the SRP/SAA pair $r=0.89$ ($p=0.0000$).

Conclusion: In the study group, all AIDs, both monogenic (FMF, TRAPS, CAPS) and sJA, were characterized by an increase in the levels of CRP and SAA, but to the greatest extent these indicators were increased in sJA and FMF, and to the least in CAPS. CRP and SAA correlate well with each other, especially with TRAPS and FMF. The maximum values of ferritin and the highest frequency of its increase were noted in the sJA group. Ferritin in the majority of pts with monogenic AIDs were normal.

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FUNCTIONAL CONDITION OF KIDNEYS IN THE LONG-TERM COURSE OF SLE IN ADOLESCENTS

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Background: Lupus nephritis is the most severe and adverse systemic lupus erythematosus (SLE) syndrome. According to modern recommendations, the clinical manifestations of active nephritis should be taken under medical control in 6 months after the start of the disease's treatment¹.

Objectives: The aim of this study was to examine the functional status of the kidneys in children with SLE in the course of the disease for more than one year.

Methods: The analysis included case histories of 43 patients with SLE, mostly females (41), aged 7 to 18 years (mean age 14.4 years) with disease duration of 4.75 ± 0.58 years of whom 22 were less than three years, 21 - more than three years. All children received corticosteroid therapy, at the time of the examination the average dose was 13.85 ± 1.86 mg per day in terms of prednisolone. The second component of therapy was azathioprine (average dose 97.61 ± 2.11 mg). All children received hydroxychloroquine (5 mg/kg per day).

To determine the functional state of the kidneys a clinical analysis of urine, a study of the scope of specific gravity of urine during the day (Zymnitsky test), the content of creatinine and urea in serum to determine the glomerular filtration rate (GFR), the level of microalbuminuria per day were evaluated.

Results: Renal involvement in the developed SLE occurred in 73.08% of patients. Among them, therapy during the first 6 months was considered quite effective in 58.06% of patients. It was found that in children with disease duration from one to three years proteinuria was registered in 68.18%, a decrease in GFR in 4.45% and hyperfiltration in 9.09%. In the group of children with duration of SLE more than three years revealed deeper changes in renal function; there was proteinuria in 90.47%, the frequency of GFR decreased was in 19.04%, a decrease of renal concentration function was in 14.28% of cases.

Indicators of renal function in children with SLE depending on the duration of the disease (M ± m)

Indicator	Duration of the disease	
	from 1 year to 3 years n = 22	over 3 years n = 21
Creatinine, mmol/l	$0,080 \pm 0,014$	$0,090 \pm 0,018$
Мочевина, mmol/l	$5,66 \pm 1,42$	$5,63 \pm 1,61$
GFR, ml/min	$117,05 \pm 19,68$	$100,20 \pm 18,98^*$
Microalbuminuria, mg/day	$24,41 \pm 13,13$	$34,73 \pm 24,76$
Density min	$1,007 \pm 0,005$	$1,006 \pm 0,005$
Density max	$1,024 \pm 0,005$	$1,019 \pm 0,005^{**}$

* $p<0,03$; ** $p<0,01$ the probability of differences when comparing between groups

Conclusion: Long-term follow-up of children with SLE over one year reveals a prolongation of renal dysfunction, which worsens after three years, and is the basis for the development of irreversible renal impairment.

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