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JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

NEUROLOGICAL INVOLVEMENT

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ABSTRACT - With the purpose of analyzing the neurological involvement due to systemic lupus erythematosus (SLE), we evaluated 17 female patients who were seen regularly at the hospital and had been diagnosed as having SLE according to classification criteria proposed by the American College of Rheumatology revised in 1982, before the age of 16. Neurological involvement was detected in 12 patients(71%): headache (35%), extrapyramidal syndrome (35%), epileptic syndrome (24%) pyramidal syndrome (24%), peripheral neuropathy (12%) and optic neuritis (6%). The findings of CT scan (58%) and cerebrospinal fluid (50%) were most closely correlated to clinical neurological involvement.

KEY WORDS: juvenile systemic lupus erythematosus, neurological involvement.

Lupus eritematoso sistêmico juvenil: comprometimento neurológico

RESUMO - Com o objetivo de analisar o comprometimento neurológico secundário ao lupus eritematoso sistêmico (LES), avaliamos pacientes que estavam em acompanhamento ambulatorial. Foram selecionadas 17 pacientes do sexo feminino cujo diagnóstico do LES foi até a idade de 16 anos, e preencheu os critérios para a classificação propostos pelo Colégio Americano de Reumatologia revisados em 1982. O comprometimento neurológico foi observado em 12 pacientes(71%): cefaléia (35%), síndrome extrapiramidal (35%), síndrome epiléptica (24%), síndrome piramidal (24%), neuropatia periférica (12%) e neurite óptica (6%). Dos exames complementares a tomografia coputadorizada de crânio (58%) e o líquido cefalorraquidiano (50%) mostraram maior correlação com a presença de comprometimento neurológico.

PALAVRAS-CHAVE: lupus eritematoso sistêmico juvenil, comprometimento neurológico.

In the literature few publications concentrate on the clinical neurological involvement in juvenile systemic lupus erythematosus (SLE). Mostly these are reports on SLE in children in whom only some neurological manifestation is described.

We performed this study in order to characterize neurological involvement due to the direct action of the juvenile SLE on central and peripheral nervous system.

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PATIENTS AND METHODS

Seventeen female patients whose SLE was identified before they were 16 years old and complied with classification criteria proposed by ACR revised in 1982[7], were submitted to a clinical neurological assessment and complementary exams: electroencephalogram (EEG); cerebrospinal fluid (CSF); cranial computerized tomography (CT); electroneuromyography (ENMG); brainstem auditory evoked potentials (BAEP) and antiphospholipidic antibody research.

The age at which the first neurologic assessment took place ranged from 3 to 18 years (mean:11,4).

In order to establish whether the illness was active or not the Urowitz et al.[8] criteria was used.

Neurological symptoms and/or abnormal clinical neurological exam and/or abnormal complementary exams allowed us to identify neurological involvement.

Patients with neurological injury secondary to the impairment of other organs and/or drug action were excluded.

RESULTS

Twelve patients (71%) had neurological involvement. Of these, 7 (58%) presented two or more different clinical neurological manifestations and 5 (42%) only one.

The relationship in time between the SLE diagnosis and the clinical neurological manifestations varied: in 1 patient (8%) the manifestations preceded the diagnosis; in 2 (17%) they were concomitant; in 7 (58%) the manifestations appeared in the course of the SLE; in 2 (17%) they were indeterminate.

The central nervous system proved to be more involved (100%) than the peripheral (25%).

The SLE was active at the time of the diagnosis of the clinical neurological manifestations in 9 patients (75%).

The clinical neurological manifestations in juvenile SLE patients are in Table 1.

The complementary exams in juvenile SLE patients are in Table 2.

Table 1. Clinical neurological manifestations in juvenile SLE patients (n=17).

Syndrome	Symptom	Sign	N	%	
Asymptomatic			5	29	
Algia	headache		6	35	
Epilepsy	focal seizure		2	12	24
	unclassified seizure		2	12	
Extrapyramidal		posture tremor	4	23	35
		tremor and cogwheel rigidity	1	6	
		chorea	1	6	
Pyramidal		hyperreflexia and clonus	3	18	24
		complete and disproportionate hemiparesis	1	6	
Optic neuritis	visual blurring		1	6	
Peripheral neuropathy	asymptomatic		1	6	12
	Pains on feet Impaired march	hypesthesia, fallen feet	1	6	

Table 2. Complementary exams in juvenile SLE patients (n=17).

Exam	Nb Patients assessed	Nb Exams with alteration	Presence neurol clinic manifest in 12 cases.	alteration
EEG	16*	2(13%)	2(17%)	cerebral suffering
CSF	16*	8(50%)	8(50%)	↑ cells-1 ↑ protein-2 ↓ glucose-3 ↑ gamma-2
CT	15*@	7(47%)	7(58%)	cerebral atrophy**
ENMG	12	2(17%)	2(17%)	signs of mononeuritis multiplex
BAEP	11	-	-	-
LA	17	1	1(8%)	positive
AAC	17	2	2(16%)	positive
VDRL	17	-	-	-

*one patient had neurological manifestation due to the malfunctioning of other organs; @ one had alteration compatible with neurocisticercosis; ** associated to hypodensity in 2 cases and to spontaneous hyperdensity of basal ganglia in 2 cases; LA, lupus anticoagulant; AAC, anticardiolipin antibody; BAEP, brainstem auditory-evoked potentials.

DISCUSSION

SLE is an autoimmune systemic chronic disease with inflammation in connective tissue and blood vessels.

The neurological involvement has not received the same attention as the other systemic manifestations because it is rarely the first manifestation. The pathophysiological mechanisms related to this involvement which can impair the peripheric or central nervous system are still questioned. Neither the primary vessels injury nor the infections, arterial hypertension nor reactions to medicine account for all the nervous system involvement.

The first article to emphasize neurological involvement in SLE children was that of Gold and Yahr[4] followed by that of Cassidy et al.[2], Yancey et al.[9], King et al.[5], and Davies and Ansell[3].

Most articles do not distinguish between neurological and psychiatric impairment labeling them neuropsychiatric disorders.

The central nervous system involvement in children has been reported as varying from 13 to 60%[1,4,6]. However in our study we found an involvement of 71%. This discrepancy may be due to the fact that we also assessed patients who had been considered neurologically asymptomatic as far as pediatricians were concerned.

The results of our study confirm the importance of neurological assessment of every child or adolescent with SLE.

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