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TROPICAL SPASTIC PARAPARESIS IN NORTHEASTERN BRAZIL

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SUMMARY — Ten possible cases of tropical spastic paraparesis (TSP) in Northeastern Brazil (Ceará) are presented. They show the typical symptoms and signs of TSP consisting of weakness of the lower limbs, spastic gait, hiperreflexia, bladder dysfunction and variable signs of posterior columns impairment. The laboratory examinations excluded other compressive, infective, degenerative or demyelinating lesions of their spinal cord. Our patients age ranged from 21 to 59 years, all were of black origin and all were of lower social class. There was a slight preponderance of females. An etiological implication of a retrovirus (HTLV-I) has been shown for TSP, but for lack of technical conditions we could not determine it in our patients, and that stands as our subsequent step in those and further cases.

Paraparesia espástica tropical no Nordeste do Brasil.

RESUMO — Os autores apresentam 10 casos de provável paraparesia espástica tropical (PET) oriundos do Nordeste do Brasil (Hospital das Clínicas — Universidade Federal do Ceará). Os sintomas consistem de fraqueza dos membros inferiores, marcha espástica, hiperreflexia, disfunção esfincteriana e sinais variados de distúrbio proprioceptivo. Outras etiologias de mielopatias foram excluídas pelos exames complementares. A idade dos pacientes ia de 21 a 59 anos, todos eram da raça negra e havia discreto predomínio de mulheres. Recentemente, tem-se atribuido ao retrovírus HTLV-I a etiologia da PET. Por falta de condições técnicas ainda não pudemos determiná-la em nossos pacientes.

Tropical myelopathies have interested clinicians and researchers since long time [14,18-20]. Those cases have been reported from different countries such as India [9,10], Seychelles Islands [8], Zaire [1], South Africa [2], Jamaica [5], Colombia [15,16,23], Martinique [21], and Peru [7]. Clinically, tropical spastic paraparesis (TSP) consists of a chronic and progressive weakness with spasticity predominantly of the lower limbs with slight impairment of the proprioception [14]. Etiologically, this disease was attributed to possibly malnutrition, toxins, treponemal agents, or parasites [13,14], and more recently to a retrovirus (HTLV-I) in the cases from Caribbean, South America and Africa [5,6,12,17,22].

From a perusal of the literature, no cases seem to have been reported from Brazil. Since, our interest in presenting some of such cases.

CASUISTICS AND METHODS

Ten patients attending to an University Hospital (Fortaleza-Ceará-Brazil) were studied and investigated along several months. Every one was hospitalized for specialized exams and after the definition of the diagnosis they have been followed up as out-patients.

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To define the diagnosis of TSP the following criteria suggested in previous papers (*16*) have been adopted: 1. No history of difficulty walking or running during school age, combined with two or more of the following items, within two years of onset: (a) increased urinary frequency, nocturia, or retention with or without penile impotence; (b) leg cramps and/or low back pain; (c) symmetrical weakness of the legs within 6 months of the onset of the disease; and (d) complaints of numbness or dysaesthesiae of legs and feet. 2. Clinical examination demonstrating increased patellar reflexes, spasticity of both legs (usually manifested by spastic gait) and absence of a sensory level, pupillary abnormalities and/or optic disc changes. 3. No history of relapses.

Besides these criteria, our TSP patients should not present fasciculations and pronounced muscle atrophy of the extremities, as well as they should have a normal myelogram and spinal fluid, the stools and rectal biopsy should not reveal **Schistosoma mansoni** and their cranio-cervical radiography should exclude basilar impression, a common pathology in our region. All patients live in the Northeast of Brazil, in the State of Ceará, that lies 4-6° south to Equator, and has a tropical oceanic climate with irregular rain periods and long periods of sunlight. This is one of the poorest areas of Brazil with a significant degree of infantile mortality.

RESULTS

The 10 patients, aged from 21 to 59 (mean of 42.3) years, were all black (10/10), predominantly females, and most of them from peasant origin (Table 1).

The principal symptoms of these patients were weakness of the legs, complaints of dysaesthesiae or numbness of the legs and feet, low back pain, increased urinary frequency and less frequently impotence and legs cramps. No history of difficulty walking or running during school age has been reported in those patients. Neurologically they presented spasticity and hiperreflexia of the lower limbs and, in some cases, of the upper limbs and a spastic gait. They had absence of a sensory level and in no case there were pupillary abnormalities, optic disc changes, history or relapses or presence of fasciculations and muscle atrophy (Table 2). Up to now their disease has an evolution from 1 to 8 years (mean: 4 years) (Table 1).

Patients		Age	Sex	Race	Origin	Profession	Time of evolution
I	JGS	59	F	Black	S. Gonçalo	—	6 years
II	AMPC	21	F	Black	S. Gonçalo	Housewife	5 years
III	PSM	54	M	Black	Fortaleza	Builder	8 years
IV	PRF	30	M	Black	Jaguaribe	Peasant	1 year
V	FCR	50	F	Black	Fortaleza	Housewife	2 years
VI	MNF	48	F	Black	Fortaleza	Dressmaker	5 years
VII	SAL	44	F	Black	Quixeramobim	Housewife	2 years
VIII	MJRS	33	F	Black	Fortaleza	Teacher	3 years
IX	RPS	49	M	Black	Aratuba	Peasant	6 years
X	MAFS	35	F	Black	Campina Grande	Housewife	2 years

Table 1 — Tropical spastic paraparesis in Northeastern Brazil: epidemiological aspects.

The different exams carried out in our patients showed a normal myelogram, a normal CSF and electrophoresis, and negative serological tests for syphilis in serum and CSF. The schistosomiasis research, performed in some of the patients, was negative. The vitamin B_{12} estimation and serum folate levels were not determined. The cranio-cervical radiograms were normal.

	Patients I	II	III	IV	V	VI	VII	VIII	IX	X	Total	%
Symptoms:												
1. History of difficulty walking or running during school age	—	—	—	—	—	—	—	—	—	—	0/10	0
2. Increased urinary frequency	—	+	—	—	—	+	+	—	+	+	5/10	50
3. Impotence	—	—	—	—	—	—	—	—	+	—	1/10	10
4. Leg cramps	—	—	—	—	+	—	—	—	—	—	1/10	10
5. Low back pain	+	—	—	—	—	—	—	—	+	—	2/10	20
6. Symmetrical weakness of the legs within six months of onset of the disease	+	—	+	+	+	+	—	+	+	+	10/10	100
7. Complaints of dysestesiae or numbness of legs or feet	+	—	—	—	+	—	—	—	—	—	2/10	20
Signs:												
1. Muscle atrophy	—	—	—	—	—	—	—	—	—	—	0/10	0
2. Fasciculations	—	—	—	—	—	—	—	—	—	—	0/10	0
3. Spasticity of lower limbs	+	+	+	+	+	+	+	+	+	+	10/10	100
4. Hyperreflexia of lower limbs	+	+	+	+	+	+	+	+	+	+	10/10	100
5. Hyperreflexia of upper limbs	+	—	—	+	—	—	+	+	—	—	4/10	40
6. Spastic gait	+	+	+	+	+	+	+	+	+	+	10/10	100
7. Presence of sensory level	—	—	—	—	—	—	—	—	—	—	0/10	0
8. Pupillary abnormalities	—	—	—	—	—	—	—	—	—	—	0/10	0
9. Optic disc changes	—	—	—	—	—	—	—	—	—	—	0/10	0
10. History of relapses	—	—	—	—	—	—	—	—	—	—	0/10	0

Table 2 — Tropical spastic paraparesis in Northeastern Brazil: clinical findings in 10 patients. (—) absence; (+) presence.

COMMENTS

The diagnosis of TSP has been suggested in those 10 cases since none of them had a familial history of difficulty running, sensory level, pupillary abnormalities and optic disc changes, fasciculations or muscle atrophy. All present a symmetrical weakness of the legs combined in a variable fashion with bladder dysfunction, low back pain and complaints of dysaesthesia of the feet and all of them had normal myelogram and spinal fluid, negative tests for syphilis and schistosomiasis and normal cranio-cervical radiograms.

All patients were of black African ancestry similar to other studies presented [12], and there was a slight preponderance of females such as in the samples of Martinique [22] and Seychelles [13]. The mean age of the patients was 42.3 years as in other series. The patients were all native and there is no history of migration. They are of lower social class and there was no report of other similar cases in their families. The ten patients possibly have subclinical malnutrition as in the general population of that social class and none of them had stigmata of severe malnutrition which could be etiologically related with their syndrome. They had not been exposed to toxins; factors such as habits, housing, occupation, farming and pet were irrelevant for their disease as compared with other neurological patients. Our cases represent a sample of continental origin unlike those of insular character described [12], but it agrees with the proximity of Ceará to the Equator, the similarity of tropical oceanic climate with long hours of sunlight and little seasonal changes. Our cases are clinically similar to those described in other parts of the world [12,14], and in none of them retrobulbar neuropathy and deafness occurred as in respectivelly 15% and 7% of the cases of Jamaica [12]. All cases evolved insidiously unlike the acute cases described in Zaire [1]. All patients presented hyperreflexia and signs of neuropathy were absent in them.

Since these patients are still alive no neuropathological examination has been performed. It is known, from some few cases studied, that the most neuropathological features of TSP are a widespread chronic meningoencephalomyelitis and the presence of demyelination, axonal loss in the posterior columns and of the pyramidal tracts [11]. Laboratory examinations did not show consistent and typical abnormalities in patients with TSP. All examinations allowed to exclude other spinal cord diseases such as multiple sclerosis, with low frequency in the tropics, tumors, infections of other origin, motor neuron disease and basilar impression. We were also aware for those myelopathies of parasitic origin as that due to *Schistosoma mansoni*. This disease is not rare in our country and is one of the important causes of myelopathies [4].

Since the report of Gessain and associates [5], the presence of anti-HTLV-I antibodies has been showed in serum and CSF of TSP patients from different countries of South America, Caribbean and Africa, so that this retrovirus outstands as an important etiological agent. Due to regional technical impossibility, we could not perform those tests in our patients and it stands as the folowing step in our research with TSP patients in Brazil, with eventual international technical cooperation.

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