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TWO BABINSKI SIGNS IN SEROPOSITIVE (HAM) AND SERONEGATIVE TROPICAL SPASTIC PARAPARESIS

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Abstract – Tropical spastic paraparesis (TSP) may or may not be associated to HTLV-I antibodies and is usually characterized by clinical and pathological spinal cord abnormalities at thoracic levels. We present here five Brazilian patients who had typical chronic idiopatic spastic paraparesis; two of them were HTLV-I seropositive (HAM) and three HTLV-I seronegative (TSP) – associated-myelopathy. Three out of these five patients also displayed clinical supraspinal involvement, indeed, platysma muscle hypotrophy or atrophy (the Babinski plus sign). These findings support the view that clinical involvement in HAM and TSP is wider than the spinal cord abnormalities usually considered. Possible non-infectious co-factors (e.g., mycotoxins) may be involved in disease pathogenesis in a multistep process of viruses, toxins and environment which may account for serological differences found in this group of patients.

KEY WORDS: HAM, TSP, Babinski plus sign, mycotoxins, neurorehabilitation.

Dos signos de Babinski en pacientes con paraparesia espástica tropical seropositiva (HAM) y seronegativa

Resumen – La paraparesia espástica tropical (PET), puede o no estar asociada con anticuerpos contra el HTLV-I y se caracteriza, usualmente, por alteraciones clínicas y patológicas a nivel de region dorso-lumbar de la medula espinal. Presentamos cinco pacientes brasileros, quienes tuvieron hallazgos típicos de paraparesia espástica crónica idiopática; dos de ellos tuvieron (HAM) y tres no tuvieron (TSP) anticuerpos, en el suero, contra el HTLV-I. En tres pacientes se encontró hipotrofia o atrofia del músculo platisma (signo de Babinski plus), demostrando que el compromiso clínico en pacientes con HAM y TSP se extiende más allá de la médula espinal torácica. Cofactores (por ejemplo, micotoxinas) podrían estar involucrados en la patogénesis de esta enfermedad, en una interacción compleja de virus, toxinas y medio ambiente, lo cual explicaría las diferencias serológicas encontradas en este grupo de pacientes.

PALABRAS CLAVES: HAM,TSP, signo de Babinski plus, neurorehabilitación.

The extension of the first toe, called the Babinski's sign is considered one of the paradigms to be evaluated in central nervous system abnormalities, and a marker of upper motor neuron involvement[1]; however, recent investigations have demonstrated that such statements are not strictly true.

In fact, the extension of the first toe is found not only in the mistakenly called upper motor neuron disorders[2] but also in lesions of the peripheral nervous system such as Guillain-Barré syndrome, several myopathies and other so-called lower motor neuron disorders[3-5]. It indicates that this reflex response is related more to a sensory-motor disintegration at central levels with a subsequent dysfunction in the motor output clinically detected as the great toe extension than to a malfunctioning of the multilevel hierarchy of the human nervous system revisited[2,6]. Further, what Joseph Babinski called the Babinski's sign was the failure of the platysma muscle to contract ipsilaterally to a hemiparesis[6]. Because of this, it was recommended to employ the term Babisnki's sign for platysma muscle abnormalities as originally described by the author and leave the term Babinski's reflex for the extension of the great toe looked for in clinical evaluations[6,7]. Likewise, if platysma muscle paresis or paralysis is present with or without neural structures abnormalities other than hemiparesis it should be named "the Babinki-plus'sign"[6].

We present here a group of Brazilian patients having chronic idiopathic spastic paraparesis with (HAM) and without HTLV-I infection (TSP) who displayed a combination of these clinical findings.

METHOD

A field study was carried out in a small community of pa-

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Table. Clinical, demographic and laboratory data.

n	G	YOB	AO	HTLV-1/2 (Serology)		PCR	Mg	MRI	Babinski's plus sign
1	M	1950	36	NR	–	–	N	No	Present
2	F	1938	34	R	+	+	N	**	Present
3	M	1950	42	NR	–	–	ND	No	Present
4	M	1955	40	NR	–	–	N	No	Absent
5	F	1939	46	R	+	ND	ND	*	Absent

n, subject number; G, gender; M, male; F, female; YOB, year of birth; AO, age, in years-old, of TSP onset; NR, not reactive; R, reactive; +, seropositive; –, seronegative; PCR, polymerase chain reaction; Mg, myelography; MRI, magnetic resonance imagin of spinal cord; N, normal; ND, not done; **atrophy; *mild atrophy.

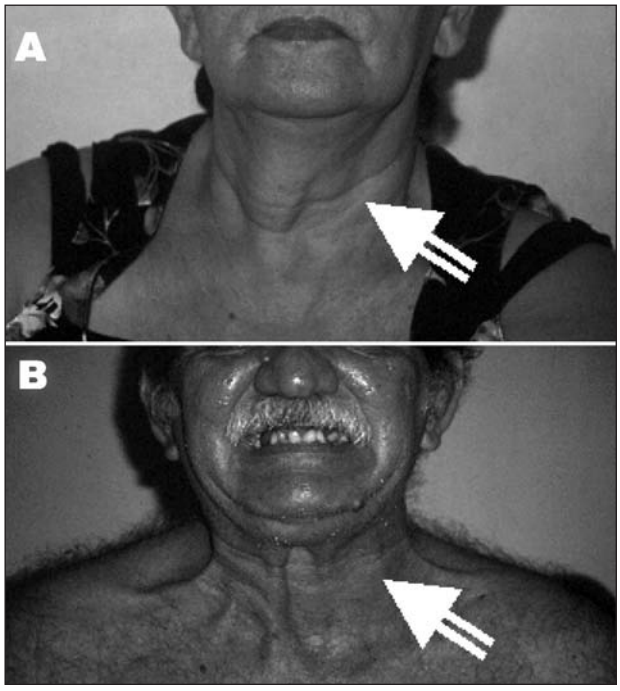

Figure. The Babinski-plus sign (green arrows). Total atrophy of left platysma muscle in a 65 years-old, female patient with HTLV-I seropositive TSP (A), and hypotrophy of the left platysma muscle in a 53 years-old, male patient with HTLV-I seronegative TSP (B). These muscle abnormalities were clearly evident while at rest (A) and during voluntary muscle effort (B).

tients with neurological disorders living at Fortaleza, Brazil (latitude 3º46' S, longitude 38º36' W, mean annual humidity 80%, and annual rainfall in between 80 to 360 mm), in 2003. We performed a throughout clinical, epidemiological and laboratory investigation in five Brazilian patients who have insidious and progressive spastic gait.

Informed consent was obtained from patients and the ethics committee of Uni.ciencias Research Group approved the study.

RESULTS

All patients had spastic paraparesis and, sometimes, cramps or progressive weakness of lower limbs, bilateral hyperreflexia and up-going toes fulfilling clinical criteria for chronic idiopatic spastic paraparesis or HTLV-I-associated myelopathy[8,9] (Table). Other clinical findings were Hoffman's sign (2), bladder disturbances (2), knees pain (1), constipation (1) and vibratory abnormalities in lower limbs (1); none of them had sensory abnormalities. Two out of these five patients (40%) were seropositive for HTLV-I antibodies (ELISA reagent confirmed by Western Blot) (HAM/TSP), and three of them (60%) displayed the Babinski's plus sign (Figure). We had not access, to perform functional neurological investigations to evaluate more accurately the supraspinal muscle-nerve involvement in these patients.

DISCUSSION

The up-going toe is not a surprising finding in this group of patients since this is a common reflex response elicited in subjects who have sensory-motor dysfunction at central levels including spasticity[6,10] one of the clinical hallmark of HAM and TSP[8,9]. What was new was to find that three of out these five patients (60%) had platysma muscle hypotrophy or atrophy called the Babinski's plus sign[6]. As far as we know this is the first time that abnormalities of this cutaneous muscle, which is innervated by some terminal branches of facial nerve[6] and high cervical cord[11], are described in clinical grounds in patients with HAM and TSP.

A recent study from Japan demonstrated abnormal magnetic resonance image lesions in the cervical cord of HTLV-I seropositive patients and it was suggested that it might be a variant of typical HAM/TSP[12]. Moreover, spinal cord abnormalities appearing in similar anatomical places like the ones described by Umehara et al.[12] also produced platysma muscle abnormalities[11] as reported in this group of Brazilian patients with HAM and TSP.

Interestingly, we and others reported some years ago, significant subclinical supraspinal abnormalities in patients with HAM/TSP, including brainstem abnormalities involving trigeminal-facial pathways recorded in the orbicularis oculi muscles after applying electrical supraorbital nerve stimulation[13,14]. These latter findings consisted of an early

contralateral reflex response, called R1k, which was found in up to 60% of patients as well as a clear sensory-motor disintegration and instability at brainstem levels[11,15] that included dysynchrony of ultranociceptive blink reflexes responses, specifically the R3 ones[11,13,16,17]. These abnormal findings seem to be due to a pathological ignition-like mechanism[16,18] as well as abnormal plastic modulation at brainstem levels.

The aforementioned explanations and findings, put together, may help to understand more clearly clinical dysfunction and aberrant plastic changes present in neuromuscular disorders associated to brainstem and upper cervical spinal cord abnormalities present in patients with spasticity regardless HTLV-I infection; and they must be considered to give appropriate credit and explain in a clearer manner original clinical signs and descriptions[6] in these days of evidence-based clinical neurology. Likewise, our results add support to the fact that the involvement in HAM/TSP is more widespread that usually thought[13-15,19], and call the attention on performing more accurate clinical evaluations to avoid misdiagnosis and delays to perform appropriate neurorehabilitation measures[6].

Lastly, the clinical picture of these small sample of patients was typical for TSP regardless HTLV-I infection[8,9,20], the latter found here in up to 40% of patients, a very close proportion to that obtained in larger seroepidemiological studies performed and reported elsewhere by different groups including the ours[8,20,21]. Environmental co-factors (e.g.,mycotoxins) including high humidity and rainfall levels[22,23] linked to disease pathogenesis would explain these uneven results and might be the hidden factors that would trigger retrovirus-associated-neurodegenerative diseases including HAM, TSP and the like.

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REFERENCES

1. Kumar SP. The Babinski sign: a critical review. J Assoc Phys India 2003; 51:53-57.
2. Carpenter R. Neurofisiología. México: Manual Moderno, 1997.
3. Gomez-Fernandez L, Calzada-Sierra DJ. Pseudobabinki. Rev Neurol 2001;32:799.
4. Leon-Sarmiento FE, Prada LJ. Babinski´s reflex, yes, Babinski´ sign, no, pseudobabinski, never!. Rev Neurol 2002;34:699.
5. Ropper AH. The Guillain-Barre syndrome. N Eng J Med 1992;326:1130-1136.
6. Leon-Sarmiento FE, Camacho JE, Bayona-Prieto J. Hemiplegia with two Babinski signs. Medicina (Buenos Aires) 2007;67:374-376.
7. Leon-Sarmiento FE, Prada LJ, Torres-Hillera M. The first sign of Babinski. Neurology 2002;59:1067.
8. Castro-Costa CM, Carton H, Santos TJ. HTLV-I negative tropical spastic paraparesis: a scientific challenge. Arq Neuropsiquiatr 2001;59:289-294.
9. Osame M. HAM: epidemiology, clinical features and pathomechanism. Gann Monograph Cancer Res 1992;39:57-68.
10. Leon-S FE, Dimitrijevic MR. Recent concepts in the pathophysiology of spasticity. Inv Clin 1997;38:155-162.
11. Ogawa Y, Sakakibara R. Platysma sign in high cervical lesion. J Neurol Neurosurg Psychiatry 2005;76:735.
12. Umehara F, Nagatomo S, Yoshishige K, et al. Chronic progressive cervical myelopathy with HTLV-I infection: variant form of HAM/TSP? Neurology 2004;63:1276-1280.
13. Leon-S FE, Arimura K, Arimura Y, Sonoda Y, Osame M. Contralateral early blink reflex in patients with HTLV-I associated myelopathy/tropical spastic paraparesis. J Neurol Sci 1995;128:51-57.
14. Yonenaga Y, Arimura K, Suehara M, Arimura Y, Osame M. Electroencephalographic abnormalities in human T-cell lymphotropic virus type I-associated myelopathy. Arch Neurol 1989;46:513-516.
15. Leon-S FE, Arimura K, Sonoda Y, Arimura Y, Osame M. Instability of R3 response of the blink reflex in patients with HAM/TSP. Funct Neurol 1994;9:199-202.
16. Leon-Sarmiento FE, Bayona-Prieto J, Gomez J. Neurophysiology of blepharospasm and multiple system atrophy: clues to its pathophysiology. Parkinson Rel Dis 2005;11:199-201.
17. Leon-S FE. Blink reflex and discomplete facial nerve palsy. Arch Med Res 2002;1:85-87
18. Devor M, Amir R, Rappaport ZH. Pathophysiology of trigeminal neuralgia: the ignition hypothesis. Clin J Pain 2002;18:4-13.
19. Cervilla J, Cartier L, García L. Brain and spinal cord magnetic resonance imaging in spastic paraparesis associated to human T-lymphotropic virus. Rev Med Chile 2006;134:1010-1018.
20. Zaninovic V, Leon-S FE. Fifteen years of follow-up on HTLV-I positive and HTLV-I negative spastic paraparesis patients in southwestern Colombia, Southamerica. J Neurovirol 1996;2:357-360.
21. Leon-S FE, Costa CM, Gaffga N. Discrepancy, coincidence or evidence in chronic idiopathic spastic paraparesis throughout the world: a meta-analysis on 2811 patients. Arq Neuropsiquiatr 1997;55:530-535.
22. Leon-S FE, Carpintero M, Gaffga N, Ocampo L, Bayona J. Mycotoxins in myelopathies of man. Lancet 1996;348:1039.
23. Leon-S FE, Zaninovic V. Geographical considerations on HAM/TSP in Japan. Rev Inst Med Trop S Paulo 1995;37:185-186.