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 idiopathic 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 región dorso-lumbar de la médula espinal. Presentamos cinco pacientes brasileños, 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; 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 but also in lesions of the peripheral nervous system such as Guillain-Barré syndrome, several myopathies and other so-called lower motor neuron disorders. 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 multiple hierarchy of the human nervous system revisited. Further, what Joseph Babinski called the Babinski’s sign was the failure of the platysma muscle to contract ipsilaterally to a hemiparesis. Because of this, it was recommended to employ the term Babinski’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. Likewise, if platysma muscle paresis or paralysis is present with or without neural structures abnormalities other than hemiparesis it should be named “the Babinski-plus’sign”.

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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Patients 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 idiopathic spastic paraparesis or HTLV-I-associated myelopathy8,9 (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 spasticity6,10 one of the clinical hallmark of HAM and TSP8,9. 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 sign6. 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 nerve6 and high cervical cord11, 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/TSP2. Moreover, spinal cord abnormalities appearing in similar anatomical places like the ones described by Umehara et al.12 also produced platysma muscle abnormalities11 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 orbiculai oculi muscles after applying electrical supraorbital nerve stimulation3,14. 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\textsuperscript{16,19} that included dysynchrony of ultranociceptive blink reflex responses, specifically the R3 ones\textsuperscript{11,13,16,17}. These abnormal findings seem to be due to a pathological ignition-like mechanism\textsuperscript{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\textsuperscript{6} in these days of evidence-based clinical neurology. Likewise, our results add support to the fact that the involvement of HAM/TSP is more widespread that usually thought\textsuperscript{13-15,19}, and call the attention on performing more accurate clinical evaluations to avoid misdiagnosis and delays to perform appropriate neurorehabilitation measures\textsuperscript{6}.

Lastly, the clinical picture of these small sample of patients was typical for TSP regardless HTLV-I infection\textsuperscript{8,9,20,21}, 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\textsuperscript{8,20,21}. Environmental co-factors (e.g., mycotoxins) including high humidity and rainfall levels\textsuperscript{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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