Comparison between the spastic paraplegia rating scale, Kurtzke scale, and Osame scale in the tropical spastic paraparesis/myelopathy associated with HTLV

Comparação entre a escala para paraplegia espástica, escala de Kurtzke e escala de Osame, na paraparesia epástica tropical/mielopatia associada ao HTLV

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ABSTRACT

Introduction: The objective of this study was to compare Osame’s scale of motor incapacity and the expanded scale of the state of incapacity of Kurtzke with the spastic paraplegia rating scale for the clinical evaluation of patients with HTLV-I-associated myelopathy/tropical spastic paraparesis (HAM/TSP). Methods: Patients with the diagnosis of infection by HTLV-I/HTLV-II and with the clinical suspicion of HAM/TSP were included in the study. Results: There were 45 patients who were evaluated. When analyzing the results of the scales, the researchers found the following average of 21.08 points for the spastic paraplegia rating scale, 4.35 points for Osame’s scale, and 4.77 points for Kurtzke’s scale. The relation between the scale of paraplegia with Osame’s was very significant with p < 0.0001, and regarding Kurtzke’s scale, there was a similar result of p < 0.0001. When comparing Osame’s, Kurtzke’s, and the spastic paraplegia rating scale with the time of disease, the researchers found a significant result of p = 0.0004 for the scale of spastic paraplegia, p = 0.0018 for Osame’s scale, and p < 0.0001 for Kurtzke’s scale. Conclusion: The spastic paraplegia rating scale has a good relation with Osame’s and Kurtzke’s scales showing a p index that is very significant that indicates that, although the scale was not initially made to be applied to patients with HAM/TSP because of the infection by HTLV, it showed to be as efficient as Osame’s and Kurtzke’s scales in evaluating the patients’ neurological conditions.

Keywords: HTLV. HAM/TSP. Osame. Kurtzke. Spasticity. Scale.

RESULTS

For were 45 patients. When analyzing the results of the scales, the researchers found the following average of 21.08 points for the spastic paraplegia rating scale, 4.35 points for Osame’s scale, and 4.77 points for Kurtzke’s scale. The relation between the scale of paraplegia with Osame’s was very significant with p < 0.0001, and regarding Kurtzke’s scale, there was a similar result of p < 0.0001. When comparing Osame’s, Kurtzke’s, and the spastic paraplegia rating scale with the time of disease, the researchers found a significant result of p = 0.0004 for the scale of spastic paraplegia, p = 0.0018 for Osame’s scale, and p < 0.0001 for Kurtzke’s scale. Conclusion: The spastic paraplegia rating scale has a good relation with Osame’s and Kurtzke’s scales showing a p index that is very significant that indicates that, although the scale was not initially made to be applied to patients with HAM/TSP because of the infection by HTLV, it showed to be as efficient as Osame’s and Kurtzke’s scales in evaluating the patients’ neurological conditions.

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CONCLUSIONS

A diagnostic criterion has not been well established yet; parameters have been described by the World Health Organization as weakness in the legs, diffused hyperreflexia, clonus, loss of vibratory sensitiveness, and urinary disturbances. Diagnosis usually requires the presence of virus or antibodies of the virus in the blood or liqueur. Osame’s scale of motor incapacity and Kurtzke’s expanded scale of the state of incapacity are used to evaluate the patients who suffer from HAM/TSP by HTLV-I; however, these scales do not demonstrate subtle improvement or worsening in the patient’s clinical condition. Schüle et al. developed a scale for the evaluation of spastic paraplegia for other types of spasticity but of easy application for patients with HTLV.
Therefore, the objective of this work was to compare Osame’s scale of motor incapacity and Kurtzke’s expanded scale of the state of incapacity with the scale for spastic paraplegia rating scale (SPRS) with the aim of evaluating the clinical situation of patients with HAM/TSP, through an instrument functionally developed for spasticity.

**METHODS**

It was a transversal study, and it was conducted through reviewing the medical promptbook that consists of a specific protocol for HAM/TSP of the patients who had consultations in the clinic of HTLV.

The patients’ promptbooks were from the clinic of the Center of HTLV from the Bahian School/Bahian Foundation for the Development of Sciences/Oswaldo Cruz Foundation. The patients had consultations between the periods of June 2008 and April 2009.

Patients included were the ones diagnosed with the infection of HTLV-I/HTLV-II through the exams ELISA and Western blot with the localization of the antibodies, anti-HTLV-I/ HTLV-II, or the localization of the virus in the blood or liquor. They must be eighteen years old or older and have the clinical suspicion of HAM/TSP after neurological evaluation.

Patients excluded from the study were those with cognitive deficit and/or psychiatric disturbance, impossibility of answering the questionnaire, other neurological diseases or any disease that can influence in answering the questionnaires, or when there is doubt about the patient’s diagnosis.

The used promptbook has a specific protocol for HTLV that contains the patient’s personal data, time of disease, medical and familiar antecedents, progression of disease, neurological exam, and laboratorial exams (viral load for HTLV, HTLV’s serotype). The specific scales that were used for the patients’ clinical evaluation were Osame’s scale of motor incapacity, Kurtzke’s expanded scale of the state of incapacity, and the SPRS.

In these cases, the analysis was evaluated through measures of frequency, averages, medians, and pattern deviations. To analyze the concordance of scales, graphs of dispersion were used to visualize the concordance between the three scales that were studied, comparing the scale of spastic paraplegia with Osame’s and Kurtzke’s. To estimate the equations of straight line that better represented the relations between the scales and to compare them with the ideal one, the simple regression analysis method was used. The set level of significance for the analysis was 5%. P values lower than 0.05 were considered statistically significant.

All participating individuals were briefed on the study and signed a free, informed consent form before the collection of samples.

**Ethical considerations**

The study was approved by the human ethics committee of the Bahia School of Medicine and Public Health.

**RESULTS**

During the period of collection, 45 patients were evaluated; among these, 30 (66.7%) were female, 15 (33.3%) were male, 43 (95.6%) were infected by HTLV-I, and 2 (4.4%) were infected by HTLV-I/II. Through the demographic characteristics, it was observed that the mean age was 52.3 years: 54.8 years for males and 50.8 years for females. Regarding the duration of the disease, the average was 8.9 years, with males having a duration of 10.5 years and females with a duration of 8.13 years.

Regarding the signs and symptoms, the researchers found the following: paraparesis in inferior members of 88.9% (40), paraparesis in superior members of 11.1% (5), hyperreflexia in superior members of 48.9% (22), hyperreflexia in inferior members of 93.3% (42), Babinski’s sign in 86.7% (39), Hoffman’s sign in 33.3% (15), Tromner’s sign in 28.9% (13), spasticity in 82.2% (37), clonus in 73.3% (33), spastic march in 86.7% (39), altered sensitivity in 8.9% (4), paresthesia in 75.6% (34), urinary disturbance in 82.2% (37), intestinal constipation in 42.2% (19), lumbar pain in 57.8% (26), and erectile dysfunction in 17.8% (8) of the patients.

When analyzing the results of the scales, the researchers found the following averages: 21.08 (±10.9) points for the scale for spastic, 4.35 (±2.15) points for Osame’s scale, and 4.77 (±1.7) points for Kurtzke’s scale. Correlation of SPRS with Osame’s and Kurtzke’s scales was also evaluated. The relationship between the scale of paraplegia and Osame’s was very significant, with p < 0.0001. For Kurtzke’s scale, the result was similar with p < 0.0001, also a significant one (Figures 1 and 2).

Comparing Osame’s and Kurtzke’s scales and the SPRS on the duration of diseases, the researchers obtained significant results, with p = 0.0004 for the scale of spastic paraplegia, p = 0.0018 for Osame’s scale, and p < 0 for Kurtzke’s scale (Figures 3, 4, and 5).
Salvador, the capital of the State of Bahia, in the northeast of Brazil, has the highest prevalence of infection by HTLV-I, when compared with other big Brazilian cities. Among blood donors in Brazil, the highest seroprevalence of bearers with anti-HTLV-I (1.5%) was detected in Salvador, State of Bahia, comparing with other capitals such as Manaus (0%), Recife (0.3%), Rio de Janeiro (0.3%), and Florianópolis (0.08%)\(^2\). The infection by HTLV-I is endemic in Japan and in many tropical countries where seroprevalence varies between 6% and 30%. Among these infected individuals, 5% to 10% usually develop not only HAM but PET as well\(^9\). Endemic cases of spastic paraparesis have been related worldwide, mainly in Japan, Malaysia, Equatorial Africa, and the Caribbean\(^17\). It is known that in endemic regions, the prevalence increases with age, and it is higher in females\(^6\). Dourado et al.\(^11\) related that seroprevalence in Salvador, because of the infection by HTLV-I, was 1.7% - 1.2% for males and 2% for females, which corroborate with the highest prevalence in female subjects. It has been related that this disease affects between 0.2% and 5% of individuals after forty years old. From the studied population, we can notice that demographic aspects, such as more women acquiring the disease, were compatible with the ones that had been identified in the clinical study by Orland et al.\(^15\), about the prevalence and clinical characteristics of the neurological disease of HTLV. However, in that study, the dominant age was determined mainly between 40 and 49 years old, which is different from what was observed in the present study where the mean age was 52.3 years. It can be related to the late diagnosis of the disease because of the difficult access of the population to specialized services or the difficulty of medical teams in identifying the disease.

The reason why 95% of the people who are infected by HTLV-I are asymptomatic is not known; the physiopathology that activates the symptomatic state and the reason for the tropism of the virus are not fully understood. Although the immune pathogenic mechanisms that are specific of PET/MAH have not been totally clarified, diverse studies have demonstrated the participation of the immune system in the development or maintenance of this neurological disorder\(^17,18\). The spastic paraparesis caused by HTLV-I is a demyelinizing chronic disease, with the presence of a slowly progressive myelopathy\(^14\). The pathogenicity of the agent is not only restricted to the pyramidal motor system; Kuate et al.\(^11\) reported two cases of cerebellar syndrome associated with an infection by HTLV-I. The incidence of HAM/TSP is lower than 1%, which suggests that for HAM/TSP to exist, there are necessary viral factors and conditions of the host\(^10,14\). Other studies show that this incidence rate of HAM/TSP is in 4-5% of the people infected by HTLV-I\(^12,13\). The main neurological findings are spastic paraparesis, lumbar pain, sensorial disturbance on inferior members, and urinary disturbance\(^11,14,22\). Despite the typical presentation of HAM/TSP, there is an increase in the number of studies that indicate an extrapyramidal pathogenic model. Subcortical and periventricular lesions of the white mass have been described in magnetic resonances of the brain. That is why some authors propose the change of the term myelopathy associated with HTLV-I to myeloneuropathy\(^2\) related to HTLV-I\(^1\). Other neurological disorders are associated with HTLV-I, as isolated peripheral neuropathy, and a condition similar to amyotrophic lateral sclerosis\(^5\). Regarding the clinical
findings in the work that characterize the tropical spastic paraparesis, a compatibility with a big study carried out by the Work Group of Infectious Disease from the Nervous System - Brazilian Academy of Neurology was observed.

The results also showed that the scale of spastic paraplegia has a good relation with Osame’s and Kurtzke’s scales, showing a very significant p value that indicates that despite not having initially applied to patients with HAM/TSP because of the infection by HTLV, it is shown to be as efficient as Osame’s and Kurtzke’s scales to evaluate the patients’ neurological condition. As there are no studies yet on the use of the scale of paraplegia for the patients with HAM/TSP, the researchers could not correlate the results with those of the other authors. Nevertheless, the study showed that its use is promising. Once the scale presents a more detailed evaluation of a patient as a whole, in this way, there is a better classification of a patient’s condition.

We can see in Figures 2 and 3 that many patients who had similar scores in Osame’s and Kurtzke’s scales had a different result in the scale for spastic paraplegia. Thus, it indicates that, probably, this last one identifies signs and symptoms that are more subtle and not observed by the first two. Regarding the duration of the diseases and the scales, the study showed that all of them have a significant relation, that is, the longer the duration of the disease, the bigger the tendency of a higher punctuation in the scales, mainly for the scale for the SPRS and Kurtzke scale. However, more studies are necessary to evaluate more details with which scale represents, with much fidelity, the duration of a disease. A scale can be more sensitive to the alterations of signs and symptoms than others. Thus, the results demonstrate that the scale of spastic paraplegia may be useful to help create a more detailed scale for another HAM/TSP occurrence. Perhaps, it can even be used in full, but further studies are needed to verify its application in patients infected by HTLV.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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