Conjugal amyotrophic lateral sclerosis

Godeiro-Junior C, Oliveira ASB, Felicio AC, Chieia MA, Gabbai AA.
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TO THE EDITOR

Sir,

With interest I read the paper of Godeiro C. et al.[1] entitled "Conjugal Amyotrophic Lateral Sclerosis".

There are several reports, as mentioned by the authors, in the literature, describing the almost simultaneously appearance of sporadic amyotrophy lateral sclerosis (SALS) in a couple, no genetically related, living together for a long time.

Of course, this circumstance strongly suggests the existence of an environmental factor triggering the disease in people, perhaps, genetically susceptible. The search for such factors has been, and it is, one of the main topics of research in SALS.

However, the particular description of Godeiro et al., as far as I can understand, does not follow the rigid clinical diagnostic concept of SALS, which requires the presence of lower and upper motor neurons clinical involvement. This happens neither with the husband of couple I, nor with the husband of family II. Both men seem to be affected by a dennervatory condition involving, apparently, just the lower motor neuron.

According to "El Escorial" criteria (1991) the isolated compromise of the spinal motor neuron reaches the 4th level of diagnostic certainty, called "suspected". The revised "El Escorial" criteria version, done later at Airlie House (1998), has positioned this criterion at the 5th level, recommending to avoid this type of patients for SALS research and for therapeutic clinical trials.

I agree with this last notion, thinking that SALS is a unique and individualized entity, which needs, to be recognized, the simultaneous clinical involvement of the cortical and spinal motor neurones. Of course, I also agree with the concept that it is a disease inhabiting the spectrum of disorders known, as a group, as "motor neuron diseases" which encompasses other clinical entities such as progressive spinal atrophy, progressive bulbar atrophy and primary lateral sclerosis.

I think that, by the time being, the illnesses embraced under the name of "motor neuron diseases" should be considered as individual disorders. This theoretical position, actually, would allow researches to focalize in one of them, when looking for its pathogenesis and aetiology.

It is my opinion that mixing the diseases could introduce bias in such type of research, delaying the finding of the cause, whose achievement might lead to a rational treatment of those conditions, a goal which, right now, seems to be very far away.

Nevertheless, I do not deny the possibility that the authors are true and what they are showing are different steps of the same process, as most probably happens with the spectrum of disorders characterized by the presence of Lewy's bodies neuronal inclusions where the compromise of autonomic ganglia, basal nuclei and cerebral cortex seems to be different stages of the same process. Despite that this could be true as well regarding motor neurone diseases, I feel that before we can admit this notion as a hard scientific proposal, we should have a marker of these illnesses able to tell us whether they are different clinical phenotypes of the same condition, a tool that is lacking yet. In the meantime, I believe that it is advisable to concentrate in what we really have right now, which only is the clinical manifestations of these disorders, avoiding any other conception which might introduce just speculative thoughts about the nature of these diseases.

REFERENCES

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Roberto E. Sica, MD, PhD
Neuroscience Department, ININCA
School of Medicine, Buenos Aires University
Paraguay 2155; piso 1. Secretaría de Ciencia y Técnica
1118 Buenos Aires. Argentina
E-mail: rsica@fmed.uba.ar

THE AUTHOR'S REPLY

We are grateful to Dr Sica for his interest in our article and for the comments and questions he raised.

We agree that not all of our patients fulfilled the El Escorial[1] clinical criteria for amyotrophic lateral sclerosis (ALS), therefore we would not be able to report their disease especifically as ALS, but only as a motor neuron disorder. In our paper, we considered as ALS those patients who had not only clinical, but also laboratory supported diagnosis of ALS.

This type of inclusion criteria may permit a misinterpretation of our data. Maybe, it would be more appropri-

ate to present the following title "Conjugal motor neuron disorder in Brazil", instead of the current one.

However, we must highlight that motor neuron disorders, especially ALS, are diseases still in search for an appropriate etiological explanation, such as many other neurodegenerative disorders (Alzheimer's and Parkinson's Disease). Different clinical presentations of ALS have been reported with a possible common etiological factor[2,3]. In presenting our article, we aimed to highlight this idea and point to the environmental factors. On the other hand, patients with the same clinical presentation may have different etiological explanation. In this point, we agree with the author comments on our paper and, while a serum biomarker is not found, we must take care and be stricter in selecting patients for clinical trials.

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Clecio Godeiro-Junior
Acary S.B. Oliveira
Andre C. Felicio
Marcos A. Chieia
Alberto A. Gabbai