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LETTER TO THE EDITOR

Sporadic adult-onset spinocerebellar ataxias

Reply: Degenerative and acquired sporadic adult-onset ataxia https://doi.org/10.1007/s10072-019-03856-w

Hélio A. Ghizoni Teive[1] · Carlos Henrique F. Camargo[2] · Renato P. Munhoz[3]

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Dear editor:

Lieto et al. [1] wrote a very interesting review article entitled "Degenerative and acquired sporadic adult onset ataxia." The authors summarized, in practical diagnostic approach, the main sporadic adult-onset ataxias including degenerative non-hereditary, hereditary, and acquired ataxias. [1] This is a very thorough review on the subject; however, we believe that there are some points that deserve further discussion.

In the group of non-hereditary degenerative ataxias, important issues could be discussed. First, in the multiple system atrophy type C, besides the classical cerebellar dysfunction, associated with dysautonomia and also pyramidal signs and parkinsonian and dystonic features, the presence of REM-sleep behavior disorder (RBD) is a very common predictive sign. [2] The RBD is strongly associated with neurodegenerative diseases, particularly synucleinopathies such as multiple system atrophy (MSA). [2] Thus, the quantification of REM sleep can be an important tool in the differential diagnosis among non-hereditary degenerative ataxias. Our group, in 2015, evaluated RBD in 50 adult patients with diagnosis of sporadic adult-onset ataxia. These patients were diagnosed with multiple system atrophy in 48% of cases and 52% received a diagnosis of idiopathic late-onset cerebellar ataxia (ILOCA) or sporadic adult-onset ataxia (SAOA). [3] RBD was diagnosed in 46% of the patients. However, among patients with MSA, RBD was found in 8.3% versus 11.5 among those with ILOCA. [3] Second, Patients with progressive supranuclear palsy (PSP) are sometimes misdiagnosed as MSA because of the presence of cerebellar ataxia. There have been reported pathologically confirmed PSP patients with predominant cerebellar ataxia (PSP-C) as their initial and principal symptom. [4] Lastly, the late-onset cerebellar atrophy of Marie-Foix-Alajouanine, a controversial entity also known as "cortical cerebellar atrophy of Marie-Foix-Alajouanine" that presents some interesting clinical features such as chorea and cognitive dysfunction, is a sub-group of ILOCA patients with very late onset (IVLOCA). [5, 6] In a Brazilian case series of 26 adult patients with ILOCA, eight of them had IVLOCA (mean age of onset was 75.5 years old). [6] Mild cognitive impairment and visual loss, due to macular degeneration, were observed in 50% of cases and chorea was found in three patients. [6]

In the group of acquired or secondary ataxias Lieto et al., [1] discussed some non-paraneoplastic autoimmune diseases, particularly coeliac disease or gluten ataxia. This entity represents a very interesting and controversial disease. Hadjivassiliou et al. [7] published in 1998 clinical, radiological, neurophysiological, and neuropathological characteristics of gluten ataxias in 28 patients. In 2017, the same research group studied the causes of progressive cerebellar ataxia, with a prospective evaluation of 1500 patients, assessed over 20 years. [8] These authors demonstrated that immune-mediated ataxias are common, and the commonest cause of sporadic ataxia was gluten ataxia (25%). [8] However, in the recently years, several other authors studied this intriguing association in sporadic and hereditary cerebellar ataxias, including spinocerebellar ataxias (SCAs), such as SCA type 2. [9, 10] In Cuban patients with SCA type 2, genetically proven antigliadin antibodies were positive in 23.4% of the ataxia patients. [9] Other interesting issue is that in our institution, with evaluation of more than 1500 patients in the last 30 years, celiac disease or gluten ataxia was not found yet (Teive et al.,

✉ Hélio A. Ghizoni Teive
hagteive@mps.com.br

[1] Movement Disorders Unit, Neurology Service, Internal Medicine Department, Hospital de Clínicas, Federal University of Paraná, Rua General Carneiro 1103/102, Centro, Curitiba, PR 80060-150, Brazil

[2] Graduate Program of Internal Medicine (Neurology), Hospital de Clínicas, Federal University of Paraná, Curitiba, PR, Brazil

[3] Movement Disorders Centre, Toronto Western Hospital, University of Toronto, Toronto, ON, Canada

unpublished data). Then, it would be very important and necessary to know the real frequency of this entity in other countries and continents.

This important review article by Lieto et al. [1] demonstrated how difficult it is to approach an adult patient with ataxia without a family history, as the differential diagnosis of sporadic ataxias is complex, and as even in tertiary centers specialized in the treatment of ataxias there are many doubts and discussions about the definitions of certain ataxias.

Authors' contributions The authors alone are responsible for the content and writing of this paper.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Lieto M, Roca A, Santorelli FM, Fico T, de Michele G, Bellofatto M, Saccà F, de Michele G, Filla A (2019) Degenerative and acquired sporadic adult onset ataxia. Neurol Sci. https://doi.org/10.1007/s10072-019-03856-w
2. St Louis EK, Boeve BF (2017) REM sleep behavior disorder: diagnosis, clinical implications, and future directions. Mayo Clin Proc 92(11):1723–1736
3. Teive HA, Arruda WO, Moro A, Moscovich M, Munhoz RP (2015) Differential diagnosis of sporadic adult-onset ataxia: the role of REM sleep behavior disorder. Parkinsonism Relat Disord 21(6): 640–643
4. Koga S, Josephs KA, Ogaki K, Labbé C, Uitti RJ, Graff-Radford N, van Gerpen JA, Cheshire WP, Aoki N, Rademakers R, Wszolek ZK, Ross OA, Dickson DW (2016) Cerebellar ataxia in progressive supranuclear palsy: an autopsy study of PSP-C. Mov Disord 31: 653–662. https://doi.org/10.1002/mds.26499
5. Fox SH, Nieves A, Bergeron C, Lang AE (2003) Pure cerebello-olivary degeneration of Marie, Foix, and Alajouanine presenting with progressive cerebellar ataxia, cognitive decline, and chorea. Mov Disord 18(12):1550–1554
6. Teive HA, Moscovich M, Moro A, Farah M, Arruda WO, Munhoz RP (2015) Idiopathic very late-onset cerebellar ataxia: a Brazilian case series. Arq Neuropsiquiatr 73(11):903–905
7. Hadjivassiliou M, Grünewald RA, Chattopadhyay AK, Davies-Jones GAB, Gibson A, Jarratt JA, Kandler RH, Lobo A, Powell T, Smith CML (1998) Clinical, radiological, neurophysiological, and neuropathological characteristics of gluten ataxia. Lancet 352(9140):1582–1585
8. Hadjivassiliou M, Martindale J, Shanmugarajah P, Grünewald RA, Sarrigiannis PG, Beauchamp N, Garrard K, Warburton R, Sanders DS, Friend D, Duty S, Taylor J, Hoggard N (2017) Causes of progressive cerebellar ataxia: prospective evaluation of 1500 patients. J Neurol Neurosurg Psychiatry 88(4):301–309
9. Almaguer-Mederos LE, Almira YR, Góngora EM et al (2008) Antigliadin antibodies in Cuban patients with spinocerebellar ataxia type 2. J Neurol Neurosurg Psychiatry 79(3):315–317
10. Lin CY, Wang MJ, Tse W, Pinotti R, Alaedini A, Green PHR, Kuo SH (2018) Serum antigliadin antibodies in cerebellar ataxias: a systematic review and meta-analysis. J Neurol Neurosurg Psychiatry 89(11):1174–1180