The shrimp sign in ataxic cerebellar progressive multifocal leukoencephalopathy

Author(s):
Francisco J Varela¹; Hernan Chaves²; Malco Rossi, MD, PhD³

Corresponding Author:
Malco Rossi, mrossi@fleni.org.ar

Affiliation Information for All Authors: 1. Departamento de Neurología, Fleni; 2. Departamento de Diagnóstico por Imágenes, Fleni. Ciudad Autónoma de Buenos Aires, Argentina; 3. Servicio de Movimientos Anormales, Departamento de Neurología, Fleni

Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.
Equal Author Contribution:

Contributions:
Malco Rossi: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
Francisco J Varela: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
Hernan Chaves: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)

Figure Count:
1

Table Count:
0

Search Terms:

Acknowledgment:

Study Funding:
The authors report no targeted funding.

Disclosure:
The authors report no relevant disclosures.

Preprint DOI:

Received Date:
2023-05-15

Accepted Date:
2023-07-26

Handling Editor Statement:
Submitted and externally peer reviewed. The handling editor was Resident and Fellow Section Editor Whitley Aamodt, MD, MPH.
An 80-year-old woman presented to the emergency department with vertigo, right hand incoordination, and postural instability five months after rituximab treatment for chronic lymphocytic leukemia. Physical examination revealed dysarthria, horizontal nystagmus, gait ataxia, and right upper and lower limb dysmetria. Brain magnetic resonance imaging showed a T2 hyperintense (Figure, A) and T1 hypointense (Figure, B) lesion in the right cerebellar white matter that demarcates the dentate nucleus (Figure, C), with pontine and middle cerebellar peduncle extension. This imaging finding corresponds to the 'shrimp sign': white matter lesion that preserves the curvilinear-shaped dentate nucleus giving a shrimp-like appearance.1 HIV serological testing was negative, and JC virus DNA was detected in the cerebrospinal fluid, establishing the diagnosis of HIV-negative progressive multifocal leukoencephalopathy (PML). Rituximab was discontinued and she showed no radiological and clinical progression after one-year follow-up. The 'shrimp sign' is a reliable indicator of infratentorial PML in patients with cerebellar ataxia.1

WNL-2023-001895_sup ---- http://links.lww.com/WNL/D86

References:
Contributions:

Francisco Varela: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data.

Hernan Chaves: Revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data.

Malco Rossi: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data.

Acknowledgment: The authors report no acknowledgment.

Study Funding: The authors report no targeted funding.

Disclosures: The authors report no disclosures relevant to the manuscript.

Research ethics and informed consent: The patient signed informed consent. Due to the case report nature of this publication and due to non-identifiable material, this manuscript was exempt from Fleni ethics board review board approval.

Figure Legend.

Non-enhanced axial brain MRI shows a shrimp-like T2 hyperintense and T1 hypointense lesion (arrows in A and B, respectively) in the right cerebellar white matter, that demarcates the dentate nucleus (delimited with a dashed line in A and B). Dentate nucleus is best identified on susceptibility-weighted images (arrowhead in C).
The shrimp sign in ataxic cerebellar progressive multifocal leukoencephalopathy
Francisco J Varela, Hernán Chaves and Malco Rossi
Neurology published online August 31, 2023
DOI 10.1212/WNL.0000000000207868

This information is current as of August 31, 2023