Teaching NeuroImages:

Medullary lesions causing dysphagia in Leigh/MELAS overlap syndrome

Bhooma R. Aravamuthan,
MD, DPhil
Audrey R. Nath, MD,
PhD
Partha S. Ghosh, MD

Correspondence to
Dr. Ghosh:
partha.ghosh@childrens.harvard.edu

Figure MRI findings of Leigh/mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) overlap syndrome

T1 (left) and fluid-attenuated inversion recovery (right) images show prior left parietal stroke characteristic of MELAS (A, B), bilateral necrotizing basal ganglia lesions (C, D), and right > left medullary lesions (E, F, arrows) characteristic of Leigh syndrome.

A 16-year-old boy with known Leigh/mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) overlap syndrome (*MTND5* T12706C mutation) developed worsening dysphagia. Examination revealed reduced palatal movements and dysarthria. MRI showed prior strokes, basal ganglia lesions, and new right > left medullary lesions (figure). He was discharged on gastrostomy tube feeds to decrease aspiration risk. Leigh syndrome is a neurodegenerative mitochondrial disorder with necrotizing basal ganglia and brainstem lesions.¹

MTND5 mutation causes rare Leigh/MELAS overlap syndrome with clinical and neuroradiologic characteristics of both.² Though dorsal medullary lesions are common in Leigh syndrome, they are rarely asymmetric or associated with dysphagia.¹

AUTHOR CONTRIBUTIONS

Bhooma R. Aravamuthan: study concept and design, critical revision of manuscript for intellectual content, acquisition of data. Audrey S. Nath: study concept and design, critical revision of manuscript for intellectual content, acquisition of data. Partha S. Ghosh: study concept and design, critical revision of manuscript for intellectual content, acquisition of data, general concept.

Download teaching slides:
Neurology.org

From Boston Children's Hospital and Harvard Medical School, MA.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES

1. Bonfante E, Koenig MK, Adejumo RB, et al. The neuroimaging of Leigh syndrome: case series and review of the literature. *Pediatr Radiol* 2016;46:443–451.
2. Taylor RW, Morris AA, Hutchinson M, Turnbull DM. Leigh disease associated with a novel mitochondrial DNA ND5 mutation. *Eur J Hum Genet* 2002;2:141–144.

Neurology®

Teaching *NeuroImages*: Medullary lesions causing dysphagia in Leigh/MELAS overlap syndrome

Bhooma R. Aravamuthan, Audrey R. Nath and Partha S. Ghosh

Neurology 2016;87:e18-e19

DOI 10.1212/WNL.0000000000002842

This information is current as of July 11, 2016

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/87/2/e18.full
Supplementary Material	Supplementary material can be found at: http://n.neurology.org/content/suppl/2016/07/11/WNL.0000000000002842.DC1
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/87/2/e18.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Clinical neurology examination http://n.neurology.org/cgi/collection/clinical_neurology_examination Mitochondrial disorders http://n.neurology.org/cgi/collection/mitochondrial_disorders Mitochondrial disorders; see Genetics/Mitochondrial disorders http://n.neurology.org/cgi/collection/mitochondrial_disorders_see_genetics-mitochondrial_disorders MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2016 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.