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

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

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.

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

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.

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
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.