# LETTER TO THE EDITOR

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### Response to Letter to the Editor

**Keywords:** Genetics - metabolic disorders, Stroke, Stroke imaging, Magnetic resonance imaging, Mitochondrial disorder

We thank Dr Finsterer for the interest in our article[1] and for providing his constructive comments.[2] We apologize for the typographic error of "metabolic" rather than "mitochondrial" in MELAS which was an obvious mistake. Of note, the acronym MELAS can actually be traced to 1984 by Pavlakis et al. who described "mitochondrial myopathy, encephalopathy, lactic acidosis, and strokelike episodes".[3] Since then, many authors have also used "encephalomyopathy" to include both encephalopathy and myopathy.[4,5]

The intent of our article was to highlight standard magnetic resonance imaging (MRI) studies as performed in an older patient with sequential stroke-like episodes (SLEs), with an eventual diagnosis of mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS). This pattern on imaging had been a clue toward the correct diagnosis, but instead the patient underwent brain biopsy, and we wished to highlight this unusual presentation so that clinicians might consider this rare condition even in older adults based on the imaging showing cortical ribboning.[1] We submitted this to the Neuroimaging Highlights section in the Canadian Journal of Neurological Sciences to serve an educational purpose in demonstrating neuroimages. Therefore, a comprehensive review of all imaging modalities used in MELAS would not be feasible, but we thank Dr Finsterer for referencing his own article expanding on this.[2] Further, a complete review of MELAS genetics was also not the focus of our brief neuroimaging highlight; although we do not disagree about its importance and appreciate the references provided in expanding on this as well.[2] In our patient, the heteroplasmy rate in blood leukocytes was 70%. No first-degree relatives had manifestations of mitochondrial disorder aside from the mother who did have diabetes mellitus.

We do not disagree with Dr Finsterer that "imaging is not carried out of SLEs but of stroke-lesions (SLLs), the morphological equivalent of a SLE on imaging".[2] Indeed, imaging is performed in patients who present with SLEs, the clinical manifestation of SLLs, and we intended for our title to reflect that imaging was acquired during these clinical events. The trigger was presumed to be focal seizure at onset of symptoms which were initially mistaken for ischemic stroke when she developed sudden onset aphasia and right hemiparesis, with no other stressors identified. We certainly also agree with Dr Finsterer that a lesion that is vascular in nature "very well follows a vascular territory".[2] In the context of MELAS, "does not follow a single vascular territory" is often understood to mean major arterial or large artery distribution, for example the middle cerebral artery, as would be seen in large vessel ischemic strokes.[6,7] This multi-territorial distribution, potentially outside of traditional vascular territories, in addition to cortical ribbon pattern, are being highlighted here with MRI to prompt the reader to consider MELAS.

Once again, we thank Dr Finsterer for his interest in our report and contributing important points of discussion that could not be covered in this short article, which was simply meant to highlight an example of images acquired during sequential stroke-like episodes in an adult patient diagnosed with MELAS, as opposed to a comprehensive review. We appreciate this additional information which further serves the educational purpose in demonstrating these neuroimages.

## Statement of Authorship

Both authors contributed equally to the letter.

## Conflicts of Interest

The authors report no disclosures.

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