Int. j. adv. multidisc. res. stud. 2022; 2(6):950-951

Received: 03-12-2022
Accepted: 10-12-2022

International Journal of Advanced Multidisciplinary Research and Studies

ISSN: 2583-049X

Letter to the Editor

Open versus percutaneous endoscopic gastrostomy in MELAS

Josef Finsterer
Neurology & Neurophysiology Center, Vienna, Austria

Corresponding Author: **Josef Finsterer**

With interest we read the article by Yamadori *et al*. on a 10 years-old female with mitochondrial encephalopathy, lactic acidosis, and stroke-like episode (MELAS) syndrome who underwent open gastrostomy because of reduced oral intake and mediation adherence due to progressive loss of motor skills and mental retardation under general anesthesia with remimazolam, fentanyl, and rocuronium [1]. MELAS manifested phenotypically with epilepsy, autism, lactic acidosis, short stature, constipation, vomiting, and Wolff-Parkinson-White (WPW) syndrome [1]. General anesthesia was carried out without major side effects and the patient profited from gastrostomy [1]. The study is appealing, but raises concerns that require further discussion.

The main limitation of the study is that heteroplasmy rates in various tissues were not provided [1]. Heteroplasmy rates of the m.3243A>G variant can strongly influence the phenotypic expression of the mutation and therefore also the reaction to remimazolam and its pharmacodynamics respectively pharmacokinetics.

We disagree with the statement that premedication was not considered because of expected difficulties regarding its administration. The patient easily received a peripheral venous catheter which is why intravenous premedication could have been easily administered as well.

Missing is the information whether the patient had myopathy with creatine-kinase (CK) elevation or not. Myopathy is a common phenotypic manifestation of MELAS [2]. Presence of myopathy can increase the risk of a malignant hyperthermia like reaction and may necessitate precautious measures.

We disagree with the notion that placement of a percutaneous endoscopic gastrostomy (PEG) requires cooperation of the patient [1]. PEG tubes are implanted in ventilated, comatose, and sedated patients. Why not in the index case? Peri-procedural risk with local anesthesia may be less than with general anesthesia for MELAS patients.

We should know why no port-A-Cath was temporarily implanted for infusions, drug administration, and blood drawings.

It is unclear whether poor seizure control was due to inadequate choice of or dosage of anti-seizure drugs (ASDs) or due to irregular administration of ASDs. This should be clarified, particularly if ASDs after surgery.

It is not comprehensible why transversus abdominis plate (TAP) block was applied [1]. The argument to reduce peri-operative opioid use is not justified as the patient received fentanyl anyway. Furthermore, the patient required only NSRI post surgery.

We disagree with the statement that MELAS is a rare condition [1]. Carrier frequency of mtDNA variants in general is 1:200 [3]. Carrier frequency of the variant m.3243A>G in the general population is estimated as 1:400 according to a UK based cohort study [4].

Overall, the study carries obvious limitations that require re-evaluation and discussion. Clarifying these shortcomings would strengthen the conclusions and could improve the study. Surgery and anesthesia in MELAS should be reduced to the absolute minimum so as not to endanger these patients.

Declarations

Funding sources: No funding was received.

Conflicts of interest: None.

Acknowledgement: None.

Ethics approval: Only secondary data were used.

Consent to participate: Not applicable.

Consent for publication: Not applicable.

Availability of data: All data are available from the corresponding author.

Code availability: Not applicable.

Author contribution: JF: design, literature search, discussion, first draft, critical comments, final approval.

Keywords: MELAS, Remimazolam, m.3243A>G, mtDNA Respiratory Chain

References

1. Yamadori Y, Yamagami Y, Matsumoto Y, Koizumi M, Nakamura A, Mizuta D, *et al*. General anesthesia with remimazolam for a pediatric patient with MELAS and recurrent epilepsy: A case report. JA Clin Rep. 2022; 8(1):75. Doi: 10.1186/s40981-022-00564-x.
2. Lorenzoni PJ, Scola RH, Kay CS, Arndt RC, Freund AA, Bruck I, *et al*. MELAS: clinical features, muscle biopsy and molecular genetics. Arq Neuropsiquiatr. 2009; 67(3A):668-76. Doi: 10.1590/s0004-282x2009000400018.
3. Wilson IJ, Carling PJ, Alston CL, Floros VI, Pyle A, Hudson G, *et al*. Mitochondrial DNA sequence characteristics modulate the size of the genetic bottleneck. Hum Mol Genet. 2016; 25(5):1031-1041. Doi: 10.1093/hmg/ddv626.
4. Nesbitt V, Pitceathly RD, Turnbull DM, Taylor RW, Sweeney MG, Mudanohwo EE, *et al*. The UK MRC Mitochondrial Disease Patient Cohort Study: clinical phenotypes associated with the m.3243A>G mutation--implications for diagnosis and management. J Neurol Neurosurg Psychiatry. 2013; 84(8):936-8. Doi: 10.1136/jnnp-2012-303528.