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## MELAS syndrome diagnosed in ICU in a 56-year-old patient presenting with status epilepticus

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Dear Editor,

The differential diagnosis of stroke in a middle-aged adult usually does not include the mitochondrial myopathy, encephalopathy, lactic acidosis and strokelike episodes syndrome (MELAS), because such a manifestation very rarely first occurs after the age of 40 [1, 2]. We report the case of a 56-year-old woman, brought to our hospital with irritability, aphasia, and ataxic gait. A brain computed tomography revealed a hypodense area on the left temporal lobe (Fig. 1). Ten hours later she developed convulsive status epilepticus and loss of consciousness. She was treated with phenytoin and transferred to the ICU, mechanically ventilated and sedated with propofol.

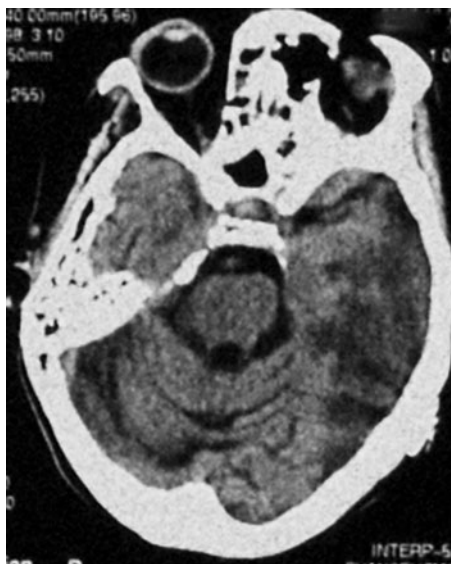
Her past medical history was marked for a hearing deficit, for which a cochlear implant had been inserted 2 years earlier, and for an episode of loss of consciousness 6 months earlier. She was functional with daily activities. The patient was childless. Three out of her four sisters had died at early age.

On ICU admission, physical examination revealed right hemiparesis. Pupils were equal and reactive

and vital signs were within normal range. Fundoscopic examination was normal. Laboratory parameters were unremarkable, with the exception of white-cell count  $18,900/\text{mm}^3$  (85 % neutrophils), aspartate aminotransferase 274 U/L, alanine aminotransferase 82 U/L, creatine kinase 9,705 U/L and lactate 3.3 mmol/L. Cerebrospinal fluid (CSF) examination was normal. The electroencephalogram showed frequent temporal spikes and slow wave complexes. Extensive imaging for brain vascular disease and laboratory investigation for autoimmune or infectious brain disease all proved negative. Magnetic resonance imaging was avoided due to the presence of the cochlear implant. On ICU day 7, the patient remained comatose. Persistent elevated blood lactate (2.5–5.3 mmol/L) was noted, despite clinical absence of seizures or haemodynamic instability. Repeat CSF revealed lactate 4.3 mmol/L, not measured in the first sample. A mitochondrial encephalopathy was then suspected. A quadriceps muscle biopsy specimen revealed “ragged red fibres”, corroborating the diagnosis of MELAS. Relatives declined further investigation

with mitochondrial DNA analysis because the result would not affect the patient’s management, and there was no reason for genetic counseling. The patient had a slow neurological improvement; she weaned from mechanical ventilation and 44 days after ICU admission she was transferred to a rehabilitation center with a Glasgow Coma Scale of 13.

First described in 1984, MELAS is a rare, maternally inherited clinical entity resulting from mutations in the mitochondrial DNA [1, 2]. The term “strokelike” refers to clinical and radiological impression of brain ischemia, albeit not conformed to a vascular territory. Although their pathogenesis remains unclear, relative deficiency of cellular adenosine triphosphate has been implicated [3]. According to diagnostic criteria, “strokelike” episodes have to appear before the age of 40 [2]. Few cases occur after the age of 50 [2–5] and none was first diagnosed in the ICU setting. Partial and complex partial status epilepticus are more commonly seen than generalized convulsive status epilepticus in MELAS [6]. ICU



**Fig. 1** Brain computed tomography scan showing a hypodense area involving the left temporal and occipital lobes

physicians must be aware of this rare entity and include MELAS in the differential diagnosis of stroke of unexplained origin, epilepsy, and elevated blood lactate levels, even in older adults.

**Conflicts of interest** On behalf of all authors, the corresponding author states that there is no conflict of interest.

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