

FDG–PET Imaging in the Investigation of Homonymous Hemianopia in a Patient With MELAS Syndrome

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Abstract: Mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome is a rare congenital disorder of mitochondrial DNA. Patients with MELAS syndrome present with a range of clinical symptoms including headaches, hearing loss, visual defects, and hemiparesis. We report a case of a previously asymptomatic 21-year-old male who presented with recurrent headache, seizures, and right homonymous hemianopia. Investigations included brain MRI, which indicated bilateral occipital white matter and cortical signal abnormality and fluorodeoxyglucose (FDG)–PET brain imaging which demonstrated markedly reduced metabolism in the occipital lobes. The patient was treated with corticosteroids and coenzyme Q and his clinical symptoms have improved.

Key Words: MELAS syndrome, FDG–PET brain, MRI brain, homonymous hemianopia, metabolism

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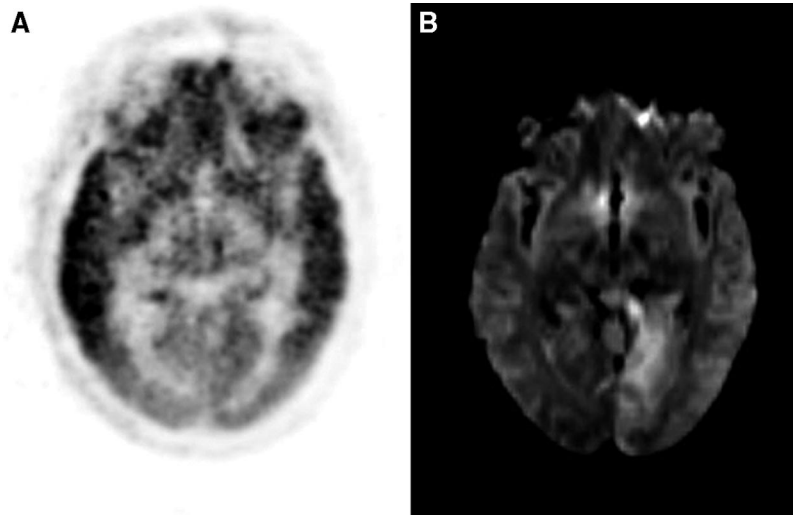


FIGURE 1. **A**, Axial FDG-PET 3D brain study on a 21 male with homonymous hemianopia demonstrates bilateral marked reduction in occipital cortex metabolism in a symmetrical distribution. There is very limited data published on the metabolic appearances of MELAS syndrome. Early reports showed a marked decrease in glucose utilization in all grey structures, with a preponderance of metabolic alterations in posterior cortical regions and thalamus, with a relative sparing of anterior cortical areas and basal ganglia.^{1,2} Cortical hypometabolism may also reflect dysfunction of associated white matter tracts. The clinical symptoms in MELAS syndrome are the result of a mitochondrial cytopathy affecting neurons or glia.² Cortical hypermetabolism has also been described³ and this may reflect early disease states where glucose uptake in the cortex is increased, but oxidative metabolism is impaired due to the abnormal function of neuronal mitochondria. **B**, Axial MR diffusion sequence (B1000) of the brain at the same level as **A** demonstrates diffuse cortical and white matter signal hyperintensity in the medial left occipital lobe.

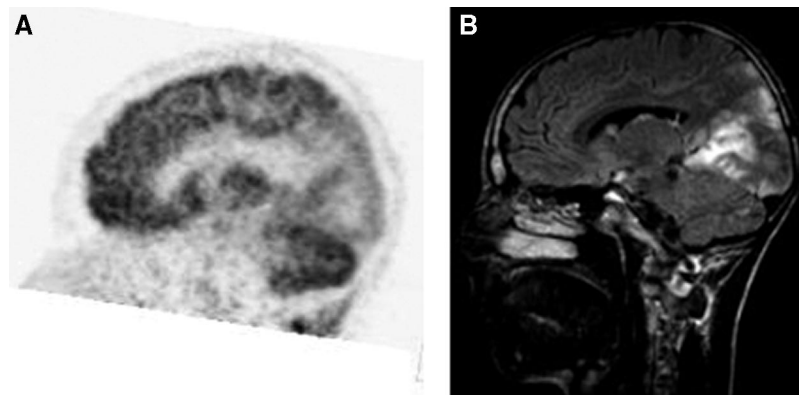


FIGURE 2. **A**, Left parasagittal FDG-PET 3D brain image demonstrates diffuse hypometabolism in the left occipital cortex, which extends into the left posterior parietal cortex. **B**, Left parasagittal FLAIR MR brain sequence at the same level demonstrates diffuse cortical and grey-white matter junction signal abnormality in the occipital lobes with an associated area of signal change in the parietal cortex superior to the parieto-occipital fissure. MRI indicates predominantly grey matter changes, but there is also a small focal area of high signal change within the posterior aspect of the left cerebellar hemisphere white matter. Of note, cerebellar metabolism at FDG-PET was normal (**A**). The most commonly described MR findings in MELAS syndrome include multiple cortical and subcortical infarct-like lesions which are not confined to the vascular territories^{4,5} and calcification of the basal ganglia.⁶ Infarct-like lesions are distributed throughout all areas of the brain, but are maximally concentrated in the occipital and parietal lobes.^{6,7} Swelling of the cortex with intracortical hyperintense bands in T₂-weighted images,⁸ representing acute laminar necrosis, has also been reported. Generalized cerebral and cerebellar atrophy is only found in severe disease.⁶