Wolff-Parkinson-White Syndrome in a Patient with Mitochondrial Myopathy

A CASE REPORT

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INTRODUCTION

- Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes syndrome (MELAS) is a maternally inherited mitochondrial myopathy.
- Cardiac manifestations are common in those with MELAS, with ~38% showing some forms of cardiomyopathy.
- Wolff-Parkinson-White (WPW) syndrome, although rare in MELAS, is the most frequently associated arrhythmia, occurring in about 13% of patients.
- We present a case where MELAS was diagnosed after WPW, highlighting the importance of considering mitochondrial myopathies in patients with WPW and implications for diagnosis and workup.

DISCUSSION

- WPW, a congenital condition characterized by intermittent tachycardia and ventricular pre-excitation arising from an accessory pathway, can manifest in those with MELAS.
- In this case, WPW syndrome and idiopathic seizures were diagnosed years prior to MELAS diagnosis.
- Individuals with MELAS commonly experience permanent multisystem impairment by adolescence or young adulthood.
- Early diagnosis of MELAS is crucial to inform families and initiate therapy (e.g., L-arginine) to reduce the chance of long-term deficits.
- A study found that 3 out of 11 pediatric patients with MELAS had WPW, indicating a potential association.
- Due to the rarity of MELAS and its association with WPW, establishing guidelines for early disease detection is questionable.

CASE SUMMARY

An 11-year-old male with multiple medical issues including seizures, vision problems, and developmental delay.

- Presenting symptoms: Worsening headaches, visual problems, nausea, and vomiting for a week.
- Diagnostic findings: CT scan with ischemic stroke concern, MRI suggested vascular abnormalities, but angiography results were inconclusive.
- Laboratory results included elevated creatinine kinase (peak 2382 U/L), hepatic aminotransferases (AST 257 U/L, ALT 96 U/L), and lactic acidosis (4.1 mmol/L).
- High suspicion for a mitochondrial disorder, specifically MELAS syndrome.
- Treatment was initiated with dextrose-containing fluids, L-carnitine, L-arginine, thiamine, riboflavin, and coenzyme Q10.
- Patient improved on the treatment and his seizure frequency decreased.
- Genetic testing confirmed a mitochondrial tRNA mutation associated with MELAS.

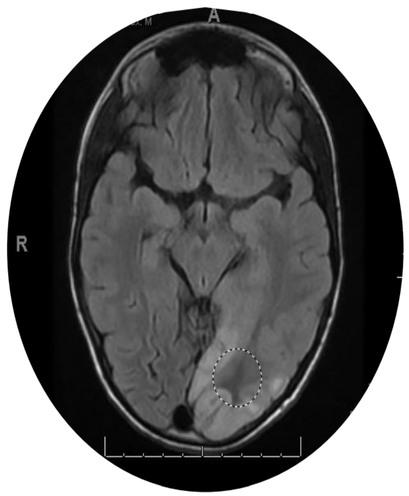

Figure 1. Diffusion restriction within the cortex and juxtacortical white matter of the left occipital lobe. Underlying T2 hypointensity within the underlying juxtacortical white matter.

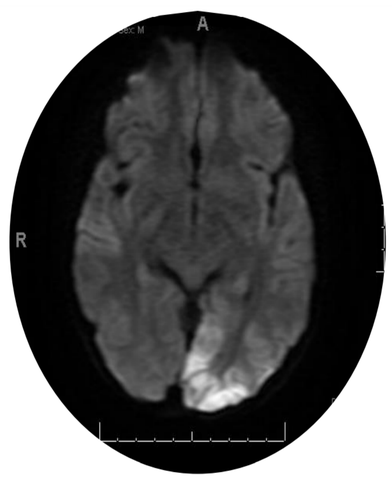

Figure 2. T2 FLAIR hyperintense edema in the left occipital lobe with underlying T2 hypointense juxtacortical white matter.

CONCLUSION

Due to significant morbidity and mortality, clinicians should screen those with otherwise unexplained neurologic and cardiac symptoms for mitochondrial myopathies, including MELAS. If negative, patients should be closely monitored for additional manifestations of underlying mitochondrial disorder, with repeat testing as indicated.

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