

Cerebellar and cortical abnormalities in paediatric opsoclonus-myoclonus syndrome

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AIM:

Paediatric opsoclonus-myoclonus syndrome (OMS) is a poorly understood condition with long-term cognitive, behavioural, and motor sequelae. Neuroimaging has indicated cerebellar atrophy in the chronic phase, but this alone may not explain the cognitive sequelae seen in many children with OMS. This study aimed to determine the extent of structural change throughout the brain that may underpin the range of clinical outcomes.

METHOD:

Nine participants with OMS (one male, eight females; mean age [SD] 14y, [6y 5mo], range 12–30y) and 10 comparison individuals (three males, seven females; mean age 12y6mo, [4y 9mo], range 10–23y) underwent magnetic resonance imaging to acquire T1-weighted structural images, diffusion-weighted images, and magnetic resonance spectroscopy scans. Neuroblastoma had been present in four participants with OMS. Voxel based morphometry was used to determine changes in grey matter volume, tract-based spatial statistics to analyze white matter integrity, and Freesurfer to analyze cortical thickness across visual and motor cortices.

RESULTS:

Whole-brain analysis indicated that cerebellar grey matter was significantly reduced in the patients with OMS, particularly in the vermis and flocculonodular lobe. A region-of-interest analysis indicated significantly lower cerebellar grey matter volume, particularly in patients with the greatest OMS scores. Diffusion-weighted images did not show effects at a whole brain level, but all major cerebellar tracts showed increased mean diffusivity when analysis was restricted to the cerebellum. Cortical thickness was reduced across the motor and visual areas in the OMS group, indicating involvement beyond the cerebellum.

INTERPRETATION:

Across individuals with OMS, there is considerable cerebellar atrophy, particularly in the vermis and flocculonodular lobes with atrophy severity associated with persistent symptomatology. Differences in cerebral cortical thickness indicate disease effects beyond the cerebellum.