Resolution of chorea movement secondary to Non-Wilsonian hepatolenticular degeneration (NWHD) post Orthotopic liver transplantation (OLT)

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Introduction

Non-Wilsonian hepatolenticular degeneration (NWHD) is a heterogeneous neurological disorder occurring secondary to chronic acquired liver disease. Genetically determined familial NWHD is rare, poorly understood, and often mistaken for Wilson's disease (WD). We present a case of a 65-year-old woman with a family history of NWHD, found to have dystonia, parkinsonism, tremor, cerebellar ataxia, progression of behavioral abnormalities who presented with cognitive decline and progression of liver failure. The patient was evaluated for and later underwent Orthotopic liver transplant (OLT).

Case Presentation

65-year-old woman with family history of early-onset cirrhosis with dystonia and dyskinesia in her father, sister, and daughter is transferred to our institution after she was noted to have accelerated progression of her neurological decline which started the year prior. The patient was not obese (BMI 27) and did not use any alcohol. Her Hgb A1C was 5.8. Huntington's disease workup was negative. Workup for causes of cirrhosis did not yield any findings including multiple 24-hour urine copper collections, and no finding of Kayser Fleischer rings on ophthalmology exam. Multiple CT and MRI brain showed linear abnormal signal foci noted along a medial portion of the bilateral lentiform nucleus in anterior to posterior orientation. The patient was diagnosed with NWHD and underwent OLT. Pathology on explanted native liver showed focal steatohepatitis, Mallory-Denk bodies, and focal mixed inflammatory infiltrates. The copper stain was negative. Post liver transplant our patient's dystonia, parkinsonism, tremor, cerebellar ataxia, and behavioral abnormalities all resolved.

Discussion

Degeneration of basal ganglia leads to movement neurological disorders. There is an association between basal ganglia-related neurological disorders and cirrhosis of the liver in the absence of acquired liver disease such as Wilson's disease. NWHD is a distinct disease entity. The disease profile of NWHD patients does not show acquired liver diseases such as alcohol or hepatitis. In NWHD specific areas of the brain, such as the basal ganglia, are more likely to be injured from liver failure. The basal ganglia is involved in the control of movement. If damage to this area is not from copper, this condition is the "non-Wilsonian" type. Non-Wilsonian hepatolenticular degeneration may represent a disorder of other poorly known toxic depositions such as manganese. We demonstrate that with liver transplantation this damage can be reversible.

Citations:

1. Mehkari Z, Mohammed L, Javed M, et al. (September 14, 2020) Manganese, a Likely Cause of 'Parkinson's in Cirrhosis', a Unique Clinical Entity of Acquired Hepatocerebral Degeneration. Cureus 12(9): e10448. doi:10.7759/cureus.10448
2. Francesca Baschieri MD,Francesca Magrinelli MD,Anna Latorre MD, PhD,Pietro Cortelli MD, PhD,Kailash P. Bhatia MD, FRCP, Movement Disorders and Liver Disease, Movement Disorders, 11 May 2021, https://doi.org/10.1002/mdc3.13238
3. Nagappa M, Sinha S, Saini JS, Kallolimath P, Singh N, Kumar A, Bindu PS, Taly AB. Non-Wilsonian hepatolenticular degeneration: Clinical and MRI observations in four families from south India. J Clin Neurosci. 2016 May;27:91-4. doi: 10.1016/j.jocn.2015.06.035. Epub 2016 Jan 4. PMID: 26765764.
4. Martinez-Ramirez D, Walker RH, Rodríguez-Violante M, Gatto EM; Rare Movement Disorders Study Group of International Parkinson's Disease. Review of Hereditary and Acquired Rare Choreas. Tremor Other Hyperkinet Mov (N Y). 2020 Aug 6;10:24. doi: 10.5334/tohm.548. PMID: 32832197; PMCID: PMC7413136.