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Case Report

MRI 'Panda Sign' in Wilson Disease: A Radiologic Hallmark of Hepatolenticular Degeneration

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Abstract

Wilson disease, or hepatolenticular degeneration, is a rare autosomal recessive disorder caused by mutations affecting copper transport, leading to copper accumulation primarily in the liver and basal ganglia. We report a case with typical neurological, hepatic, and ophthalmologic features, emphasizing the radiological hallmark signs and diagnostic approach. The discussion highlights key imaging findings, differential diagnoses, and current therapeutic strategies.

Keywords: *Wilson Disease; Hepatolenticular Degeneration; Giant Panda Sign*

Abbreviations

MRI: Magnetic Resonance Imaging; DWI: Diffusion-Weighted Imaging; FLAIR: Fluid-Attenuated Inversion Recovery; SWI: Susceptibility-Weighted Imaging; AST: Aspartate Aminotransferase; ALT: Alanine Aminotransferase

Introduction

Wilson disease, or hepatolenticular degeneration, is a rare autosomal recessive disorder caused by mutations affecting copper transport, leading to copper accumulation primarily in the liver and basal ganglia.

Case Report

A 13-year-old boy presented with progressive tremor, gait instability, dysarthria, and behavioral changes including depression and personality alterations. Neurological examination revealed mask-like facies, spasticity, and rigidity. There were no obvious abdominal complaints at the initial presentation.

Brain MRI demonstrated bilateral symmetric T2/FLAIR hyperintensity in the putamina and other deep gray nuclei, with the classic "face of the giant panda" sign - preserved signal in the red nuclei and lateral substantia nigra pars reticulata, hyperintense tegmentum, and hypointense superior colliculus. Diffusion-weighted imaging showed restricted diffusion, suggesting active lesions in the early phase.

These striking MRI findings prompted further evaluation for metabolic and genetic disorders. Laboratory investigations revealed elevated AST and ALT, hyperbilirubinemia, markedly increased urinary copper excretion, and low serum ceruloplasmin. Clinical

reassessment identified hepatosplenomegaly with subtle stigmata of chronic liver disease. Slit-lamp examination confirmed Kayser-Fleischer rings.

Liver biopsy confirmed chronic hepatic changes compatible with Wilson disease. This case highlights the importance of recognizing characteristic neuroimaging patterns that can lead to an early diagnosis, even before overt hepatic symptoms become evident.

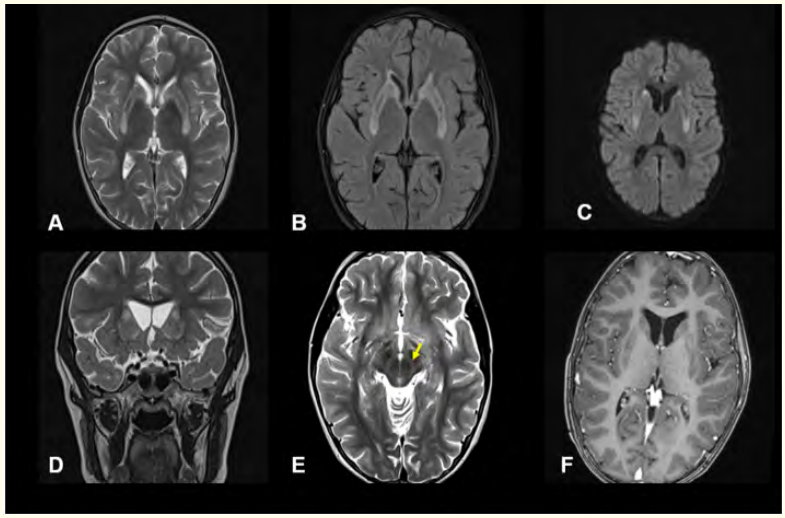

Figure 1: *MRI of the brain: Axial T2-weighted image (A) and coronal FLAIR image (B) show bilateral hyperintensity of the caudate nuclei and putamina. Diffusion-weighted imaging (C) demonstrates restricted diffusion, indicating active lesions. Axial T2-weighted image (E) reveals the characteristic "face of the giant panda" sign in the midbrain, with preserved signal in the red nuclei and high signal in the tegmentum (arrow). No post-contrast enhancement is observed following gadolinium administration (F).*

Discussion

Wilson disease results from a defect in the ATP7B gene, leading to impaired copper incorporation into ceruloplasmin and defective biliary excretion. Excess copper accumulates in multiple organs, notably the liver and brain, causing a broad clinical spectrum [1]. Neurologically, basal ganglia involvement is typical, with imaging findings often being bilateral and symmetric. The "face of the giant panda" sign is a highly suggestive neuroradiological marker [2].

Differential diagnoses in pediatric bilateral basal ganglia lesions include inherited metabolic disorders such as Leigh syndrome, Huntington disease, and pantothenate kinase-associated neurodegeneration (PKAN); acquired metabolic/toxic insults such as hypoglycemia, hyperglycemia, chronic hepatic encephalopathy, manganese toxicity, and bilirubin encephalopathy; and vascular or hypoxic-ischemic injury [3].

The mainstay of treatment is life-long administration of copper-chelating agents to maintain negative copper balance, along with dietary copper restriction. Liver transplantation remains the definitive treatment for patients with acute liver failure or decompensated cirrhosis unresponsive to medical therapy.

Conclusion

Early recognition of Wilson disease is vital. MRI, especially the "giant panda sign," provides a strong diagnostic clue and, when combined with biochemical and ophthalmologic findings, enables timely treatment to prevent irreversible neurological and hepatic damage.

Conflict of Interest

Declare if any financial interest or any conflict of interest exists.

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