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Images of Interest / Imagens de Interesse

The Giant Panda Sign on MRI as a Neuroimaging Hallmark of Wilson Disease

O Sinal do Panda Gigante na Ressonância Magnética como Marco Neuroimagimológico da Doença de Wilson

Rafael Caldas Prata Matos[1], Julia Morales[1], Maria Beatriz César[1], Marcelo de Queiroz Pereira da Silva[2], Márcio Luís Duarte[3], Leonardo Furtado Freitas[4]

[1]Faculdade de Ciências Médicas de Santos, Santos (SP), Brazil
[2]WEBIMAGEM TELERRADIOLOGIA, São Paulo (SP), Brazil
[3]Universidade de Ribeirão Preto - Campus Guarujá, Guarujá (SP), Brazil. Diagnósticos da América S.A., São Paulo (SP), Brazil. Instituto de Ensino e Pesquisa DASA (IEPD), São Paulo(SP), Brazil
[4]Florida International University - Herbert Wertheim College of Medicine, Baptist Health of South Florida and Radiology Associates of South Florida, Miami (FL), USA.

Address

Márcio Luís Duarte
Department of Radiology
Universidade de Ribeirão Preto (UNAERP) – Campus Guarujá
Av. D. Pedro I, 3.300, Enseada, Guarujá-SP, Brazil. ZIP CODE: 11440-003
e-mail: marcioluisduarte@gmail.com

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Abstract

Wilson disease (WD) is a rare genetic disorder characterized by pathological copper accumulation, primarily affecting the liver, central nervous system, and other organs. It commonly presents in young individuals with hepatic, neurological, and psychiatric manifestations. Neurological involvement results from copper deposition in the basal ganglia and brainstem and can be detected by magnetic resonance imaging (MRI), which may demonstrate characteristic findings such as the giant panda sign. Diagnostic suspicion is supported by laboratory abnormalities, including elevated urinary copper excretion, increased free serum copper, and altered liver enzymes. WD may mimic other liver diseases and neurological conditions, making an integrated clinical, laboratory, and imaging-based approach essential. Treatment relies on copper-chelating agents, such as penicillamine, or zinc therapy, combined with dietary copper restriction. Early diagnosis and prompt initiation of therapy are crucial to prevent disease progression and to improve long-term outcomes.

Keywords

Magnetic resonance imaging; Liver cirrhosis; Wilson disease.

Resumo

A doença de Wilson (DW) é uma condição genética rara caracterizada pelo acúmulo patológico de cobre, acometendo principalmente fígado, sistema nervoso central e outros órgãos. Manifesta-se com quadros hepáticos, neurológicos e psiquiátricos, sendo mais comum em indivíduos jovens. As alterações neurológicas decorrem do depósito de cobre nos gânglios da base e tronco encefálico, podendo ser identificadas pela ressonância magnética (RM), que revela sinais característicos como o sinal do panda gigante (giant panda sign). A suspeita diagnóstica é reforçada por achados laboratoriais, incluindo elevação do cobre urinário e do cobre livre sérico, além de alterações das enzimas hepáticas. A DW pode mimetizar outras hepatopatias e doenças neurológicas, exigindo abordagem diagnóstica integrada. O tratamento baseia-se em agentes quelantes de cobre, como penicilamina, ou em sais de zinco, associados a medidas dietéticas. O diagnóstico e tratamento precoces são fundamentais para prevenir progressão da doença e melhorar o prognóstico.

Palavras-chave

Imagem por ressonância magnética; Cirrose hepática; Doença de Wilson.

Case Presentation

A 20-year-old man with a previous diagnosis of liver cirrhosis presented with a seven-month history of lower limb edema. The patient also reported experiencing spontaneous episodes of dizziness and denied any mood swings in the last few months. He denied any history of alcohol or tobacco use and had received the hepatitis B vaccine. The patient's medical history includes a surgical intervention for intestinal perforation approximately four years prior. Physical examination revealed significant edema in the lower limbs bilaterally, ascites, and skin pallor. Simultaneously, complementary diagnostic procedures revealed elevated levels of serum free copper and urinary copper, in addition to elevated transaminases (ALT, AST) and gamma-glutamyltransferase (GGT). Brain magnetic resonance imaging (MRI) revealed bilateral symmetric hyperintense signal abnormalities involving the caudate nuclei and putamina on axial FLAIR images (Figure 1A), in addition to the characteristic giant panda sign in the midbrain

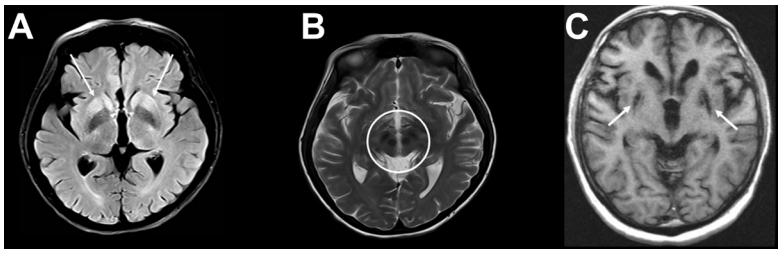


Figure 1 – (A) Axial FLAIR image demonstrating bilateral symmetric hyperintensity involving the caudate nuclei and putamina (white arrows), consistent with edema and gliosis secondary to copper deposition. (B) Axial T2-weighted image demonstrating the classic "giant panda sign" in the midbrain (white circle), characterized by preserved signal intensity of the red nuclei and lateral portions of the substantia nigra surrounded by hyperintensity of the tegmentum. (C) Axial T1-weighted image demonstrating bilateral hyperintensity of the lentiform nuclei (white arrows), associated with volume loss of these structures.

(Figure 1B). The diagnosis of Wilson disease (WD) was confirmed based on laboratory and imaging findings. Then treatment was initiated with zinc sulfate.

Discussion

WD is a genetic disorder characterized by copper accumulation in the liver, brain, kidneys, and corneas. It manifests primarily through hepatic, neurological, and psychiatric symptoms.[1] Typically diagnosed in individuals aged 5–35, the condition's prevalence is about 13.9 per 100,000 people, with higher rates in consanguineous populations and among Asians.[2]

WD is caused by mutations in the ATP7B gene, leading to impaired copper excretion and its accumulation in tissues. Symptoms typically affect the liver, central nervous system, and other organs.[1] Liver involvement can present as chronic hepatitis or cirrhosis, and neurological symptoms arise from copper accumulation in the basal ganglia and brainstem, causing ataxia, dysarthria, and tremors. Psychiatric symptoms may include emotional lability and depression, while the Kayser-Fleischer ring is a common but not pathognomonic ophthalmological finding.[2]

This case's skin pallor may be linked to anemia, probably associated with liver failure.[2] Laboratory tests, including urinary copper measurement, are crucial for diagnosing WD. Copper excretion greater than 100 μg in 24 hours in adults suggests the disease. Although total serum copper levels reflect both bound and free copper, measuring the free copper fraction is more diagnostic but lacks standardized methods.[1]

Liver dysfunction is common in WD, often presenting with abnormal liver tests, cirrhosis, or acute liver failure. Biochemical markers like ALT and AST levels can indicate hepatocellular damage, as seen in this case.[1] Neuroimaging, particularly MRI, is recommended in suspected WD, as it may demonstrate abnormal signal changes in the basal ganglia, thalami, and brainstem, some of which may improve after treatment. Genetic testing and liver biopsy can also aid in the diagnosis of WD.[2]

WD shares biochemical and histological features with other liver diseases, such as non-alcoholic fatty liver disease and autoimmune hepatitis. The basal ganglia are also affected by other systemic conditions, including neurodegenerative diseases and infections.[3]

Early treatment with copper-chelating agents, such as D-penicillamine or trientine, is essential to improve outcomes. Zinc therapy and dietary changes help achieve negative copper balance. Liver transplantation may be necessary for patients with acute liver failure or terminal cirrhosis.[1]

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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