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NEUROLOGICAL PICTURE.....

Faces of the giant panda and her cub: MRI correlates of Wilson's disease

A 26 year old man with cirrhosis and subacute cognitive decline was admitted with a decrease in his activities of daily living. Hyperpigmentation of his lower extremities, asterixis, and Kayser-Fleischer rings were observed. Neurologic examination revealed psychomotor retardation, upper extremity weakness, hypertonia, a left upper extremity tremor, and extensor plantar responses. Serum ceruloplasmin and urine copper studies confirmed the diagnosis of Wilson's disease. He subsequently developed stereotypical episodes of fever, diaphoresis, tonic decorticate posturing, and autonomic instability followed by somnolence. Infectious investigation and serial EEG were unrevealing. Progressive neurological decline ensued, despite D-penicillamine, trientine, and

zinc sulphate, leading to akimetic mutism and rigidity with profound autonomic dysfunction.

Serial MRI revealed symmetric bilateral areas of T2-hyperintensity involving subcortical white matter, basal ganglia, external capsules, thalami, midbrain, and pons. Progressive involvement of the midbrain demonstrated the characteristic MRI evolution of the "face of the giant panda" (Figure 1, A, C, and E).¹ Contemporaneous evolution of dorsal pontine signal abnormalities (Figure 1, B, D, and F) resembled the face of a cub, with eyes formed from the central tegmental tracts. Encircling signal abnormality was possibly because of involvement of the superior cerebellar peduncles, pedunculopontine tegmental nuclei, rubrospinal tracts, or lateral lemnisci.² Selective vulnerability

and progressive involvement of midbrain and dorsal pontine structures may be chronicled on MRI by the faces of the giant panda and her cub, respectively.

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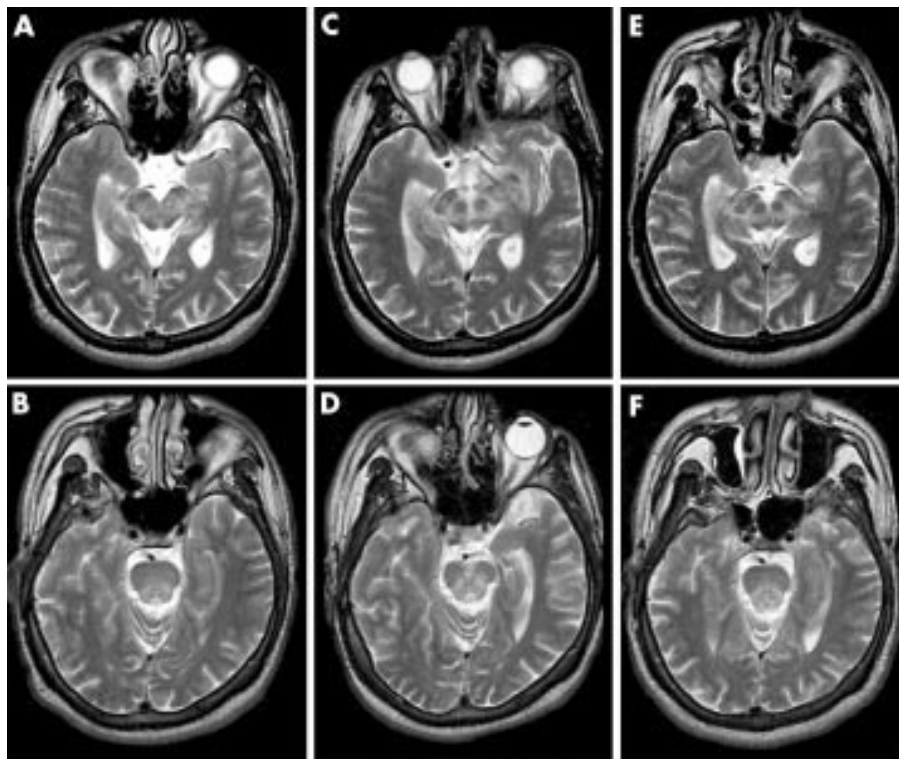


Figure 1 Serial T2-weighted MRI of Wilson's disease over a six month period illustrating evolution of the characteristic "face of the giant panda" in the midbrain (A, C, and E) and face of her cub in the dorsal pons (B, D, and F).