

Face of the Teddy Bear in Wilson's Disease: More Than Just One Kind of Beast in the Midbrain

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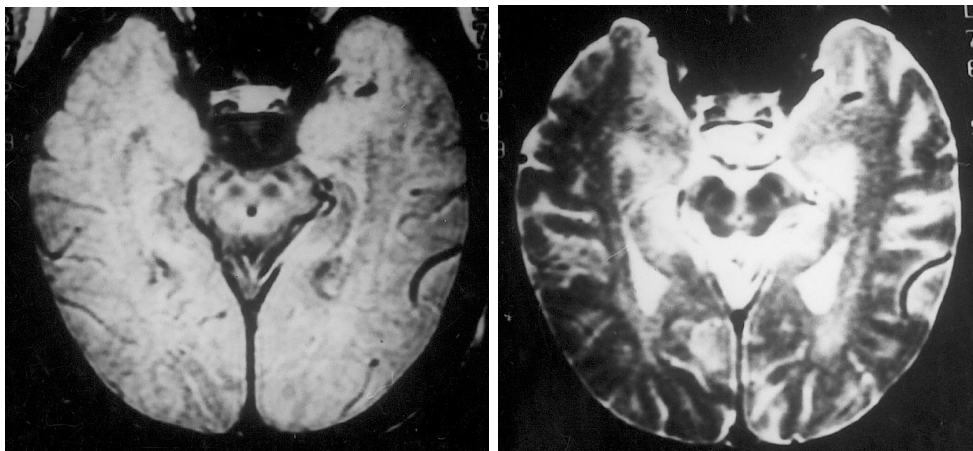


Figure 1. MRI of a 30-year-old man who had been diagnosed WD for 14 years. He presented mild parkinsonism with hand tremor as chief complaint. On both T2-weighted (right) and proton (left) images, the midbrain reveals "face of the teddy bear".

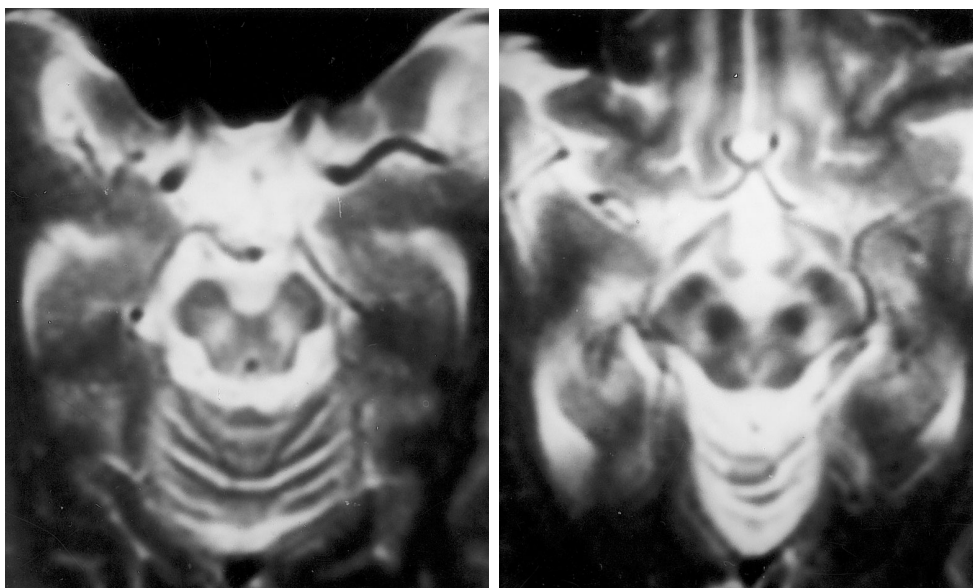


Figure 2. MRI of a 37-year-old man who had WD for 17 years. Neurological presentation was predominantly dystonia and dysarthria. Again, the appearance of the midbrain on T2-weighted image resembles typical "face of the teddy bear", but that on T1-weighted image reveals "face of the bat".

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Wilson's disease (WD) is an autosomal recessive disorder of copper metabolism affecting mainly the liver, the cornea, and the brain⁽¹⁾. Neurological manifestations are predominantly dystonia or parkinsonism⁽²⁾. On CT or MRI, lesions are commonly seen in the striatum, particularly the putamen^(3,4). Midbrain atrophy is also common. T2-weighted MRI images of the midbrain may reveal a characteristic "face of the giant panda", consisting of high signal intensity in the tegmentum except for the red nucleus, preservation of signal intensity of the lateral portion of the pars reticularis of the substantia nigra, and hypointensity of the superior colliculus⁽⁵⁾.

We report two patients with WD who had faces of other beasts on the MRI images of the midbrain. The diagnosis of WD was based on neurological presentation, family history, presence of Kayser-Fleischer corneal rings by the slit-lamp examination, low blood ceruloplasmin level, excessive urinary excretion of copper, presence of liver cirrhosis detected by abdominal echography, and/or liver biopsy.

Their MRIs revealed striatal lesions, especially in the putamen, cerebral and brainstem atrophy, and thalamic lesion in one case. In addition, their midbrains all showed "face of the teddy bear" on T2-weighted image (Figs. 1 and 2) and "face of the bat" on T1-weighted image in one case (Fig. 2).

The appearance of certain animal faces in WD is due

to pathological involvement of the red nucleus, substantia nigra, and to a lesser degree the dorsal tegmentum⁽²⁾. In our experience, the characteristic appearance of the midbrain on T2-weighted image resembles more the "face of the teddy bear" than the "face of the giant panda".

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