


Kayser-Fleischer corneal ring

Josef G. Heckmann, MD, PhD, Christoph J.G. Lang, MD, PhD, Bernhard Neundörfer, MD, PhD, Michael Küchle, MD, PhD, Erlangen, Germany

Editor’s Note: This same NeuroImage was previously printed in black and white (Neurology 2000;54:1839). Because this figure is so instructive, it is being reprinted in color for the benefit of our readers.

A 26-year-old man was diagnosed with Wilson’s disease in 1981. d-Penicillamine treatment was started but discontinued because of increased tremor. We first treated the patient in our ICU in 1987 for acute neurologic deterioration after mild brain injury caused by a fall. On admission, he was stuporous and unable to communicate verbally. We observed vertical gaze palsy, an increase in muscle tone, and a prominent circular Kayser-Fleischer corneal ring (figure, A). The laboratory findings revealed low serum copper (400 μg/L), low ceruloplasmin (7 mg/dL), and elevated 24-hour urine copper (1403 μg/1800 mL urine/day). Treatment with d-penicillamine was reinstated, along with physiotherapy and ergotherapy. The patient, now 44, is ambulatory and was recently readmitted for follow-up. Mild dysarthria, mild bilateral dysdiadochokinesia, and mildly elevated muscle tone of all four limbs were found, and the Kayser-Fleischer corneal ring had markedly regressed (figure, B). Serum copper (203 μg/L) and ceruloplasmin (3.9 mg/dL) were diminished. d-Penicillamine will be continued.

The presence of a Kayser-Fleischer corneal ring1,2 may correlate with treatment and markedly regress after successful decoppering.

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