

**46th ANNUAL DIAGNOSTIC SLIDE SESSION, 2005
REFERENCES AND DIAGNOSES**

MODERATOR: E. Tessa Hedley-Whyte, M.D.

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Case 2005-2

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Diagnosis: Hepatolenticular Degeneration (Wilson's Disease)

Comment: After a clinical diagnosis of Wilson's disease was entertained, an examination disclosed Kayser-Fleischer rings in the cornea. The 24 hour urine copper excretion was 74 μ g, which is elevated, but not greatly. The middle third of the putamen on each side had necrosis. Alzheimer type I astrocytes were tentatively identified, in addition to the readily demonstrable Alzheimer type II neurons. A rhodamine stain for copper was negative in brain sections. An alternative diagnosis offered at the session, prior to the presenter's discussion, was aceruloplasminemia.

References:

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Mercer JF: The molecular basis of copper-transport disease. Trends Mol Med 2001; 7:64-69.

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Strausak D, Mercer JF, Dieter HH, et al: Copper in disorders with neurological symptoms: Alzheimer's disease, Menkes, and Wilson's disease. Brain Res Bull 2001; 55:175-185.