

Kayser-Fleischer Ring

A 10-year-old male presented with inability to walk alone due to weakness of both upper and lower limbs for one year along with slurred speech and drooling of saliva for three years. There was no history of convulsion, jaundice, measles or recent vaccination. He was emotionally labile and had slurred speech. There was hypertonicity of all four limbs with dystonia, bilateral extensor plantar reflex, ataxia and intention tremor. Liver was just palpable. Examination of eyes revealed presence of Kayser-Fleischer (KF) ring (*Fig. 1*) bilaterally. On slit lamp examination, the ring (3 mm in width) appeared as a golden-brown deposit at the level of the Descemet's membrane of cornea (*Fig. 2*). A diagnosis of Wilson's disease was made. The serum ceruloplasmin was 3.5 mg/dL, 24 hours urinary copper was 120 µg/dL. The patient was treated with d-penicillamine and the KF ring disappeared.

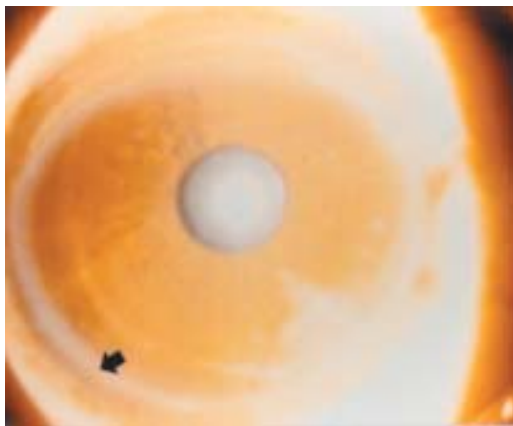


Fig. 1. Kayser-Fleischer ring in cornea.

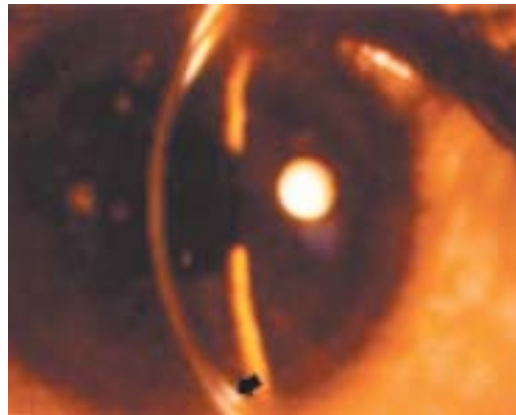


Fig. 2. Slit lamp examination reveals golden brown deposit in Descemet's membrane.

KF ring is the pathognomic sign of Wilson's disease. It may be absent in young patients with hepatic symptoms but is always present in patients with neurological symptoms. It is present in 98% of neurologically symptomatic and 40% of asymptomatic patients with Wilson's disease. The ring appears as a golden-brown to green deposit at the level of the Descemet's membrane of cornea. It ranges from 1 to 3 mm in width, progressing centrally. It is usually bilateral. Deposition begins in the superior cornea, then the inferior cornea, and eventually forms a complete ring. Treatment with d-penicillamine causes the ring to disappear, in the reverse order of its formation.

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