

NEUROLOGICAL PICTURE

Sunflower cataract in Wilson's disease

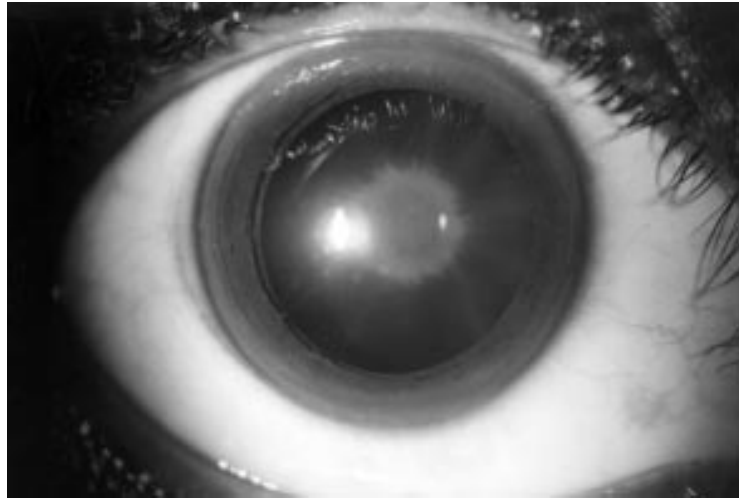


Figure 1 Right eye showing Kayser-Fleischer ring and sunflower cataract.

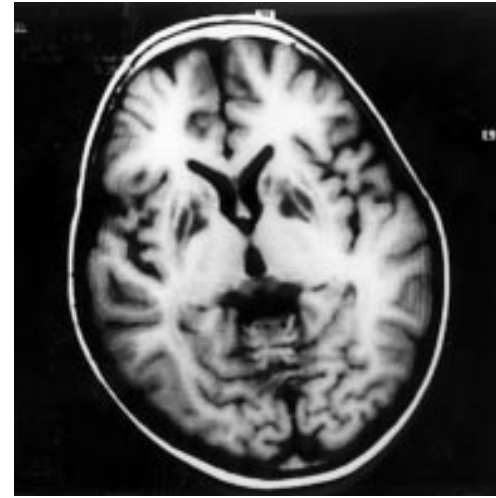


Figure 2 Brain MRI showing a bilateral hypointense signal in the putamen on T1 weighted image.

Wilson's disease is an autosomal recessive, multisystem disorder due to impaired hepatic excretion of copper. There is excessive accumulation of copper in the CNS, liver, kidneys, cornea, and other organs.¹ This accumulating copper leads primarily to damage of the liver and brain. Predominant presentation includes dystonias, tremor, dysphasia, dysarthria, gait and limb ataxia, and neuropsychiatric manifestations. The Kayser-Fleischer ring, a brownish coloration of the outer margin of the cornea in Descemet's membrane, is diagnostic of Wilson's disease, which regresses on treatment with metal chelating drugs.^{2,3} Sunflower cataract was first described in Wilson's disease by Seimerling and Oloff.⁴ We report a case with classic sunflower cataract with bilateral putaminal hypodensities on CT and hyperintense signal on T2 weighted image on MRI.

This 10 year old girl had progressive visual loss for the past 2.5 years and recurrent jaundice for the past 2 years. She had had progressive difficulty in writing for 2 months with tremors and abnormal posturing of her right arm. She also had dysphasia and dysarthria of the same

duration and was practically mute, but with preserved comprehension. On examination, she had bilateral sunflower cataract and a Kayser-Fleischer ring (fig 1). Her serum copper concentration was 180 µg/dl (normal 14–140 µg/dl) and her ceruloplasmin concentration was 1.4 mg/dl (normal 2–17 mg/dl). Brain MRI showed a hyperintense signal bilaterally in the basal ganglia on T2 weighted image (fig 2).

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- 2 Finelli PF. The Kayser-Fleischer ring: hepatolenticular degeneration (Wilson's disease). *Neurol* 1995;45:1261–2.
- 3 Johnson RD, Campell RJ. Wilson's disease: electron microscopy, x ray energy spectroscopic and atomic absorption spectroscopic studies of corneal copper deposition and distribution. *Lab Invest* 1982;46:546–69.
- 4 Siemerling E, Oloff H. Pseudosklerose (Westphal-Strumpell) mit cornealring (Kayser-Fleischer) and doppelseitiger Scheinkatarakt, die nur bei seitlicher Beleuchtung sichtbar ist und die der noch Verletzene durch Kupfersplitter entstehenden Katarakt ähnlich ist. *Klin Wochenschr* 1922;1:1087–9.