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LETTER TO EDITOR (VIEWERS CHOICE)

KAYSER-FLEISCHER RING IN WILSON DISEASE: UTILITY OF ANTERIOR SEGMENT OPTICAL COHERENCE TOMOGRAPHY

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Wilson's disease (WD) is a hepatolenticular degeneration caused by mutations in the ATP7B gene. It is an autosomal recessive disorder resulting in excessive copper deposition, particularly in liver, eyes, and brain. This occurs due to a deficiency of ceruloplasmin, a copper binding protein, often leading to liver cirrhosis. A characteristic ocular sign of Wilson disease is the Kayser-Fleischer (KF) ring, represent copper deposition in Descemet's membrane of the cornea.

KF ring is most common in neuro WD, detected in 78–85% of cases. Its prevalence in hepatic WD ranges from 36-62%, while it may be detected in 10% of asymptomatic patients.[1] Traditionally KF rings are detected using slit-lamp biomicroscopy. However anterior segment optical coherence tomography (AS-OCT) has emerged as a promising tool, offering a non-invasive, objective, high-resolution cross-sectional imaging tool that can detect subtle KF ring especially in early stage or uncooperative patients

Challenges with slit lamp examination
Copper deposits primarily in the anterior chamber angle at Schwalbe's line within the Descemet's membrane. Early detection with a standard slit-lamp is challenging since the angle view is obscured by the corneal limbus. Copper deposits only become visible after crossing the limbus.

Role of AS-OCT
AS-OCT is a high-resolution non-contact optical imaging technique using infrared light (wavelength 1310 nm) permitting greater penetration, allowing better visualisation of the structures at the angle. The KF ring appears as a distinct hyperreflective band at the level of the Descemet's membrane on AS-OCT. It displays greenish-yellow to brownish coloration on the imaging scale, signifying copper accumulation at the corneal periphery. AS-OCT captures high-resolution, reproducible images without requiring corneal anaesthesia, with each scan taking less than 20 seconds per eye. This makes it particularly advantageous for patients with neurological symptoms such as tremors or dystonia or in early stage, enabling early diagnosis and treatment before significant copper deposition occurs.[2]

Clinical Comparison of AS-OCT and Slit-Lamp Examination

- Several studies support the higher sensitivity of AS-OCT:
- Sridhar et al. (2017) reported 100% detection by both methods in a small cohort, but AS-OCT provided clearer demarcation and quantification.[3]
- Broniek-Kowalik et al. (2019) demonstrated AS-OCT detected KF rings in 86% of WD cases versus 58% by slit-lamp.[4]
- Ormeci et al. (2022) showed that AS-OCT identified rings in a significantly higher proportion of patients across all clinical subtypes, especially in hepatic forms where slit-lamp detection was <10%.[5]

These findings underscore AS-OCT role in identifying subtle rings and its utility in difficult-to-examine populations.

Conclusion
AS-OCT is a promising diagnostic and monitoring tool for Kayser-Fleischer rings in Wilson disease. Its advantages—non-invasiveness, rapid imaging, high sensitivity, and objective quantification—make it particularly useful in pediatric patients and those with limited cooperation. It complements slit-lamp examination, especially when initial findings are inconclusive or minimal. Incorporating AS-OCT into routine diagnostic protocols may enable earlier detection and treatment of Wilson disease.

Key message

- AS-OCT is a highly sensitive, non-invasive tool for detecting KF rings.
- Particularly useful in children and neurologically impaired patients.
- AS-OCT provides objective documentation and follow-up, unlike the subjective slit-lamp exam.

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