Suspecting Wilson's Disease: Guide Your Ophthalmologist

DESCRIPTION

A 15-year-old male, a diagnosed case of Wilson's disease, presented to ophthalmology out-patient department for examination of Kayser-Fleischer rings (K-F rings). He had a Child-Pugh class A chronic liver disease complicated by portal hypertension with grade 1 esophageal varices. His systemic and neurological examination was within normal limits.

Ophthalmological examination (diffuse illumination, 60° oblique) with completely exposed corneas, showed diffuse, golden-brown crescentic deposit at superior and inferior quadrant of corneas (Figure 1a). The slit-lamp examination revealed the fine, powdery dispersion of deposits to be present at the Descemet's membrane (DM) of corneas (Figure 1b). The gonioscopy showed a broad, golden-brown crescentic band which sharply ends at the Schwalbe's line (Figure 1c). The Schwalbe's line represents the termination of corneal endothelium, the target site of the copper deposition in Wilson's disease. These ring-shaped deposits are termed as K-F rings.

Kayser-Fleischer rings are one of the diagnostic signs of Wilson's disease. In chronic cases, a careful diffuse light examination in a semi-dark room may reveal these golden-brown, crescentic, corneal deposits. The whole cornea should be exposed for better superior and inferior corneal examination. The gonioscopy may provide broader and clearer picture of these DM-endothelial corneal deposits, especially in early phase of Wilson's disease.

Current knowledge:

- Kayser-Fleischer rings are golden-brown deposits of copper at the level of corneal Descemet's membrane-endothelium.
- The presence of K-F rings is a classical feature in diagnosis of Wilson's disease.
- The routine slit-lamp biomicroscopy does not always detect K-F rings.

Highlights of our description:

- Complete exposure of cornea reveals the superior limbus normally covered by upper eyelid.
- The gonioscopy should be done especially in the cases where no K-F ring is visible on routine slit-lamp biomicroscopy.

Figure 1 Ophthalmological examination (diffuse illumination, 60° oblique).

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- In early stages of Wilson's disease, the gastroenterologist/hepatologist must emphasize the need of gonioscopy for revealing the corneal deposits.

CONFLICTS OF INTEREST

The authors have none to declare.

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