Commentary: Kayser-Fleischer-like rings in patients with hepatic disease

This issue of the Indian Journal of Ophthalmology features an interesting article evaluating the association of bilirubin level and Kayser-Fleischer-like ring in patients with hepatic disease.[1]

A common reason for ophthalmology referral from the gastroenterology department is to identify Kayser-Fleischer (KF) ring in presumed cases of Wilson's disease. KF ring is a golden brown pigmented deposition of copper and copper chelates deep in the cornea in the Descemet's membrane which was first described by Kayser in 1902 and by Fleischer in 1912.[2] Wilson reported patients of familial hepatic cirrhosis with progressive lenticular degeneration in 1912.[2] The pigmentation of the Descemet's membrane in such cases was described separately by Siemerling and Orloff, and Jess in 1922.[2] Harry and Tripathi in 1970, described the electron microscopic appearance of the KF ring as electron-dense deposits of copper of varying sizes lying mainly in the Descemet's membrane.[3] Uzman and Jakus[4] also confirmed the presence of copper in the KF ring by the histochemical and electronic microscopic study.

KF rings were considered pathognomonic of Wilson's disease till Fleming *et al*.[5] observed pigmented rings in patients with primary biliary cirrhosis and Jones[6] noted similar corneal ring in a patient with intrahepatic cholestasis. While corneal copper levels could not be measured, there was a high copper content in the serum and liver in these patients leading to the conclusion that KF rings may be indicative of high copper levels in the body.[6]

However, Frommer *et al*.[7] in 1977 reported three patients with normal serum ceruloplasmin and copper levels who had corneal rings clinically similar to early KF ring. The diagnosis of these cases included active chronic hepatitis, cryptogenic cirrhosis, and neonatal hepatitis. Though hepatic copper concentration and urinary copper excretion rates were mildly raised, the levels did not reach the range seen in symptomatic Wilson's disease.

Pseudo Kayser – Fleischer rings or K-F like rings like those described in the article in IJO[1] are supposedly present in the corneal stroma rather than the Descemet's membrane. The authors correlate these rings with higher serum bilirubin levels.[1] Such rings were first described by Nagral and colleagues[8] who proposed that such rings could be bile pigment rings. They also suggested the differentiating points between the KF ring and KF like ring. However, the images provided with the paper of Nagral and colleagues suggest the location of the pigment ring is at the deep cornea or Descemet's membrane and not stroma. Thus, confirmation of the actual presence and characterization of bile pigment rings, if any may need further evaluation along with histopathological analysis.

In conclusion, the presence of K-F rings was considered to clinch the diagnosis of Wilson's disease. But, they are also seen in patients who have unexplained central nervous system disease, abnormal serum bilirubin levels, cirrhosis of the liver, rickets, renal tubular acidosis, and unexplained Coomb's negative hemolytic anemia.[9] Finding KF rings may not be the "eureka " moment for the diagnosis of Wilson's disease anymore.[9]

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