A 24-year-old male since birth presented with low vision. The patient has experienced significant complications related to the disease, including liver failure and hearing loss. Best-corrected visual acuities were no light perception OD and 20/100 OS. Open-angle glaucoma was present bilaterally with severe superior and inferior visual defects in the left eye. The right eye had a dense intumescent cataract, limiting posterior segment examination in the right eye. In the left eye, vitreous opacities and multiple discrete hypopigmented spots sparing the fovea were noted. The hypopigmented spots correlated with preretinal hyperreflective bodies seen on optical coherence tomography. this patient had an ultra- widefield fundus appearance with accompanying ocular coherence tomography scan showing more extensive deposits.