A 50-year-old Iranian diabetic woman with night blindness was referred to our clinic. Complete ophthalmic examination including ophthalmoscopy after dark adaptation and paraclinical evaluations such as fluorescein angiography and electroretinography were performed. Retinal neovascularization and preretinal haemorrhages compatible with high-risk characteristic proliferative diabetic retinopathy were observed bilaterally. In addition, a golden-yellowish discoloration of posterior pole was noted in her both eyes. The diagnosis of Oguchi's disease was made when this discoloration disappeared after dark adaptation for 3 h. Electroretinograms also confirmed the diagnosis by showing a slow negative wave followed by a slow positive wave in the photopic condition and absent a- and b-waves in the scotopic state. We concluded that proliferative diabetic retinopathy might occur in a patient with Oguchi's disease. This report represents this association in an Iranian patient for the first time.