A Case of Oguchi Disease with Diabetic Retinopathy

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Abstract

Purpose: To report an Iranian patient with diagnosis of Oguchi disease associated with diabetic retinopathy.

Methods: A 50-year-old 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.

Results: In the both eyes, retinal neovascularization and preretinal hemorrhages compatible with high-risk characteristic proliferative diabetic retinopathy were observed. In addition, a golden yellowish discoloration of posterior pole was noted in her both eyes. The diagnosis of Oguchi disease was made when this discoloration disappeared after dark adaptation for 3 hours. 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.

Conclusion: Proliferative diabetic retinopathy may occur in a patient with Oguchi disease. This report represents this association in an Iranian patient for the first time.

Keywords: Oguchi Disease, Diabetic Retinopathy, Night Blindness, Retinal Neovascularization, Preretinal Hemorrhages, Electroretinograms


Introduction

Oguchi disease, a recessive form of congenital stationary night-blinding disorders, is characterized by typical golden metallic color of fundus (Oguchi, 1907). This peculiar yellowish iridescent sheen disappears after prolonged dark adaptation (Mizuo-Nakamura phenomenon).1,2

It is believed that Oguchi disease affects only the eyes and no other systemic association has been reported for this disorder. On the other hand, most of the cases have been seen in Japan and Pakistan and as we are aware, this rare disease has not been previously reported from Iran.1-4 In this report, an Iranian patient with Oguchi disease and the association of this disorder with diabetes mellitus and diabetic retinopathy are presented.

Case Report

A 50-year-old woman was referred for ophthalmic examination by an endocrinologist. Having history of IDDM (Insulin Dependent Diabetes Mellitus) for 15 years, she complained of recent blurred vision and mild night blindness.
Complete ophthalmologic examination including funduscopy after dark adaptation and paraclinic evaluations such as fluorescein angiography and electroretinography were performed. Institutional review board approval was obtained from the local ethics committee and certified by the Vice-chancellor for research affair of the university.

Best corrected visual acuity was 20/30 in her both eyes. Fine rubeosis iridis was detected in the eyes. Intraocular pressures were normal. Fundus examination of both eyes revealed golden-yellowish color of the posterior poles together with retinal neovascularization and preretal hemorrhage (Figure 1 A and B).

Oguchi disease with high-risk characteristic stage of proliferative diabetic retinopathy was suspected; therefore, the eyes were patched for 3 hours. The golden-yellowish color was disappeared (Mizuo-Nakamura phenomenon) (Figure 1 C and D).

Fluorescein angiograms confirmed the diagnosis of high-risk characteristic proliferative diabetic retinopathy. Electroretinograms demonstrated slow negative waves followed by slow positive waves in the photopic condition. Both waves were nearly undetectable in the dark-adapted (scotopic) state (Figure 2). This pattern was not changed after 3 hours patching. Panretinal photocoagulation was performed for her diabetic proliferative retinopathy.

Figure 1. A and B: Fundus photographs in normal illumination, C and D: after 3 hours eye patching

Figure 2. A and B: photopic electroretinograms, C and D: scotopic electroretinograms
Discussion
This case represented an \textit{Iranian} diabetic patient with Oguchi disease who developed high-risk characteristic diabetic retinopathy. Oguchi disease is a rare disorder that has been previously reported from \textit{Japan} and \textit{Pakistan}.\textsuperscript{1-4} We believe that this case is the first report of Oguchi disease from \textit{Iran}.

Oguchi disease is a recessively inherited form of stationary night blindness with Mizuo-Nakamura phenomenon.\textsuperscript{5} These patients adapt very slowly to the dark, but their rhodopsin regeneration is normal. The physiologic defect appears to be in the retinal circuitry rather than the visual pigments. Once these patients are dark adapted, just a brief flash of light (too short to bleach the visual pigments) can destroy their dark sensitivity.\textsuperscript{6}

\textit{Bergmsa} et al reported an 18-year-old asian american man who was evaluated for nonprogressive nyctalopia since birth. His visual acuities with minus-ten-diopter glasses were $20/20$.\textsuperscript{7} Our case also had good visual acuities in spite of having proliferative diabetic retinopathy.

\textit{Hirose} et al first described the pattern of electroretinograms in two cases with Oguchi disease. They demonstrated a slow negative wave followed by a slow positive wave in the electroretinograms of his patients.\textsuperscript{8} In electroretinograms of our case, we found similar findings in photopic condition. In scotopic state, however, a- and b-waves were not detectable. Francois also reported a case whose electroretinogram showed absent scotopic components.\textsuperscript{9} \textit{Nagata} found a transient appearance of a scotopic b-wave after long period of dark adaptation.\textsuperscript{10}

Conclusion
Considering the prevalence of diabetes mellitus, occurrence of diabetic retinopathy in a patient with Oguchi disease could be accidental. However, since this association had not been reported in the literature, we presented our patient as the first case having both diabetic retinopathy and Oguchi disease concurrently.

References