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Darkness leading to diagnosis

Case

A 42-year-old gentleman presented to the hospital for regular ophthalmic examination. His best-corrected visual acuity was 6/6 in both eyes (OU). The anterior segment examination was normal. Dilated fundus examination revealed a golden metallic sheen in the retina OU [Fig. 1a]. The optic disc and macula were normal. Detailed history revealed that the patient had difficulty in driving at night with defective night vision and was normal during daytime. There was no similar family history.

What is Your Next Step?

A. Subject the patient to dark adaptation and repeat fundus examination.
B. Electroretinography
C. Ophthalmic examination of family members
D. All of the above

Findings and Management

The patient was subjected to 45 min of dark adaptation. The golden metallic sheen disappeared and the fundus regained normal color following the dark adaptation, suggestive of the Mizuo–Nakamura phenomenon [Fig. 1b]. The metallic sheen reappeared in the fundus following re-exposure to light for 30 min. Electroretinogram (ERG) of both eyes showed reduced rod response with normal cone function. Family members were screened and genetic counseling was given. The patient was advised to avoid night driving and the need for regular follow-up.

Diagnosis

Oguchi's disease

Correct answer: D. All of the above

Discussion

Oguchi's disease is a rare autosomal recessive form of congenital stationary night blindness. It is associated with mutations in the genes for arrestin (SAG) or rhodopsin kinase (GRK1), both of which are important in rod phototransduction.[1] The Mizuo–Nakamura phenomenon is characterized by a golden or gray-white metallic sheen in the fundus which disappears after prolonged dark adaptation and is presumably due to an excess of extracellular potassium in the retina because of a reduced potassium scavenging capacity in retinal Muller cells.[2] Electroretinogram (ERG) shows a characteristic undetectable dim flash rod response. Strong flash ERG shows marked a-wave reduction and may have a low b: a ratio. Light-adapted ERG is normal. After prolonged dark adaptation (1–3 h), there is recovery of rod-mediated response but to only the first bright flash.[3] No treatment is available so far for Oguchi's disease; thus, night vision aids can be suggested for patients with night blindness. As most patients are used to their slow dark adaptation since birth, they may never complain and thus the condition may remain undetected. In patients with night blindness, Oguchi's disease should be ruled out with detailed clinical evaluation and diagnostic tests.

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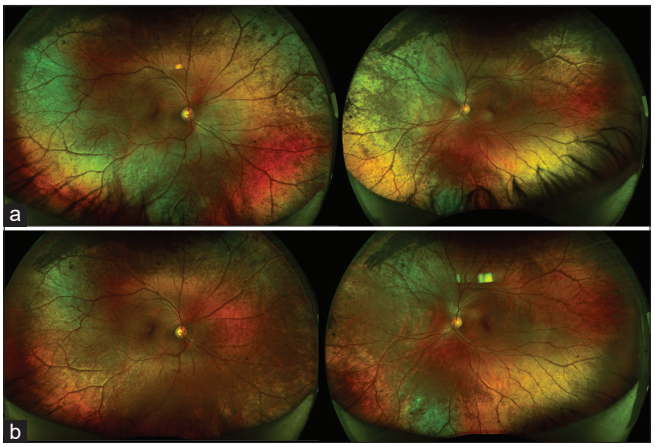

Figure 1: (a) Fundus photograph of both eyes showing golden metallic sheen of the retina before dark adaptation. (b) Fundus photograph of both eyes after 30 min of dark adaptation showing disappearance of metallic sheen exhibiting Mizua–Nakamura phenomenon

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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