Local macular ERG in patients with Best's disease

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Abstract. We examined a pair of siblings, a 10-year-old girl (case 1) and a 12-year-old girl (case 2), with Best's disease. The visual acuity was nearly normal in both patients. The central visual field measured with auto-plot tangent screen revealed a small relative paracentral scotoma only in the left eye of case 2. We failed to detect any abnormality in photopic and scotopic electroretinogram (ERG) recorded with Ganzfeld stimuli and the electrooculogram light rise was absent in both patients.

Local macular ERG was recorded under a fundus monitor by infrared television fundus camera with test spots of 5, 10, and 15 degrees in diameter. The center of the stimulus spot was always on the fovea during the recording. The local macular ERG was absent in both patients. Our results of local macular ERG may indicate disturbance of the central portion of the retina.

Introduction

Best (1905) described a family in which eight of 59 members were affected with orange-red lesions in the macular. Since his description, this relatively rare condition has been known as Best's disease and much has been reported about the affliction.

The clinical features of Best's disease have been well documented; markedly abnormal electrooculograms (EOG), normal mass electroretinogram (ERG), characteristic fundus appearance, and dominant inheritance are important findings to the diagnosis. Peripheral visual field, dark adaptation, and color vision in early stages have been reported normal (Deutman, 1971; Krill et al., 1966). Normal macular ERG has also been reported in a few patients in early stages of this disease (Bierdorf and Diller, 1969; Deutmann, 1971).

We report two siblings in early stages of this disease because of a previously unreported feature: non-recordable local macular ERG.

Case reports and methods

Case 1 is a 12 year-old girl. She was first seen by an ophthalmologist in 1979. Visual acuity at that time was 0.9 in the right eye and 1.0 in the left eye. Intermittent exotropia was observed. Macular lesion was noted in both eyes.
Case 2 is the 8 year-old sister of Case 1. She was first seen in 1981 together with Case 1. Macular lesion was also observed in both eyes. The family history revealed that only the two sisters were affected, while their one brother was normal. Both parents and their immediate families had a negative history of eye problems.

We first examined the patients in 1984 after they failed a school vision test. Visual acuity of Case 1 was 0.9 in the right eye and 0.8 in the left eye. Visual acuity of Case 2 was 1.0 in the right eye and 1.0 in the left eye. Auto-plot tangent screen showed a small relative paracentral scotoma only in the left eye of Case 1. In both patients, the fundus showed a scrambled egg appearance in the macula. Fluorescein angiogram showed hyperfluorescence in the macular lesions (Figure 1).

Mass ERGs were recorded with a corneal contact lens electrode (Life-Teck ERG-jet) after pupillary dilation with a combination of 0.5% phenylephrine hydrochloride and 0.5% tropicamide. The time constant and high cut frequency of the preamplifier (Nihon-Kohden AB-620G) were 0.1 sec and