Case Report

Electrophysiological Eye Examination Changes in a Patient with Sjogren's Syndrome

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Abstract

Sjogren's syndrome is one of the most common autoimmune diseases. It may exist as either a primary syndrome or as a secondary syndrome associated with other autoimmune diseases such as rheumatoid arthritis and systemic sclerosis. Patient with Sjogren's syndrome have certain visual system involvements with dry eye being the most common type. These patients may also exhibit certain pathological changes in their retina and visual pathway.

Here we report the electrophysiological recordings including visual evoked potential, electroretinography and electrooculography findings in a patient with Sjogren's syndrome.

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Introduction

Sjogren's syndrome (SS) is an autoimmune disease. In this disease the immune system attacks tears and saliva glands causing dry eyes and dry mouth [1]. While SS was once considered a rare disease, it is now considered the second most common autoimmune disease after rheumatoid arthritis. Its prevalence is estimated at 1 % (0.1-4.8 %), with an incidence of 7 per 100,000 in the United States [1]. The true prevalence of SS is hard to determine because of the frequent changes in diagnostic criteria. Variation in the method of diagnosis might be caused by the training of the clinician making the diagnosis (ophthalmologist, oral surgeon, rheumatologist, internist) and inclusion of markers that are potentially more sensitive. Visual system is among the organs which might be involved in patients with SS. The principal ocular manifestation is decreased tear production leading to chronic irritation [2]. Along with decreased tear production and subsequent corneal damage these patients may also exhibit some pathological changes in their retina.

Jack et al., [3] in 2015 performed a study on retina of nine SS patients (5 male, 4 females; age range 3-23 years). All patients exhibited photophobia and macular crystals. Retinal pigment epithelium (RPE) atrophy was present in 10 of 18 eyes. Also the full retinal thickness was reduced by 22 %, the inner nuclear layer was reduced by 30 % and the outer nuclear layer was reduced by 40 % [3]. This thinning suggests that there is outer retinal injury in SS, which could be the result of direct toxicity, loss of trophic factors, or altered development. Dysfunction of various sensory pathways may be also present in SS patients [4,7].

Hydroxychloroquine (HCQ) is frequently used to treat primary SS [8], and the incidence of retinopathy among patients using this antimalarial medication is relatively high [9]. Therefore, electrophysiological examination of visual system including electroretinography (ERG), electrooculography (EOG) and visual evoked potential (VEP) are performed in SS patients prior to HCQ administration and the use of HCQ should be avoided in presence of retinopathy.

Here we report the electrophysiological recordings including visual evoked potential, electroretinography and electrooculography findings in a patient with Sjogren's syndrome.

Case report

A fifty-six years old lady with SS was referred to Basir Eye Clinic, Tehran, Iran, for electroretinography and electrooculography examination. She claimed that her disease was not diagnosed in time and after several examinations of her organs, finally a dentist suspected the Sjogren's syndrome due to certain characteristic changes of her teeth. Her dentist referred the patient to a rheumatologist for further evaluation and the rheumatologist confirmed the diagnosis of SS. She had also consulted with an ophthalmologist several times due to dry eyes, for which artificial tears had been prescribed.

Her best corrected visual acuity at admission was 10/10 and she had normal fundus. The rheumatologist had decided to start hydroxychloroquine (HCQ) treatment, for which ocular examination was necessary, so the patient was referred to our center.

In our center the ophthalmologist ordered ERG and EOG examinations. The electrophysiologist decided to perform visual evoked potential (VEP) exam beside these two ordered examinations. Both ERG and EOG were subnormal, but the VEP findings were normal. Regarding the ERG, b wave amplitude was reduced to less than 50% of

normal value (Right eye voltage 45 μv, left eye voltage 32 μv). The normal value for ERG b wave amplitude is above 100 μv. Regarding the latency of b wave in ERG, the values obtained were in normal range (Right eye 42 μv, left eye 44 μv). In EOG the Arden Index (AI) values obtained were 1.62 and 1.38 for right and left eyes respectively. The normal value for AI is 1.80 to 2.80.

Discussion

The ERG and EOG were subnormal in our patient. It is a well-known fact that the retina of SS patients might be affected, and ERG and EOG can screen for the degeneration produced in different layers of retina. Changes in different layers of retina have been previously reported among SS patients [3,10]. In a study by Willemsen et al., [11] including fifteen SS patients no changes in EOG and ERG were reported, which is in contradiction with our findings.

Another electrophysiological technique which was used in our evaluation was visual evoked potential (VEP). The result was normal and the visual pathway was not affected in our patient. Sekaric et al., [12] have reported a 52-year-old woman suffering from SS with normal VEP, which is similar to our results, but in a study reporting an Indian family in 2009, the two siblings with SS were found to have abnormal visual evoked potentials [7].

Conclusion

Retinopathy and visual pathway disturbances among SS patients are relatively frequent, therefore evaluation of electrophysiological findings before treatment initiation and close follow up monitoring in patients receiving hydroxychloroquine is recommended.

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Footnotes and Financial Disclosures

Conflict of interest:

The authors have no conflict of interest with the subject matter of the present study.