

Case Report

Electrophysiological Eye Examination of a Patient Suffering from Tuberous Sclerosis and Probable Retinal Dystrophy

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Abstract

A twelve-year-old girl suffering from tuberous sclerosis complex was referred to Basir Eye Clinic, Tehran, Iran, for electrophysiological eye examinations, including visual evoked potential (VEP) and electroretinography (ERG). The patient's medical history showed that she experienced seizures at the age of ten, for which treatment with vigabatrin was initiated. Her field of vision was abnormal, potentially due to the side effects of vigabatrin. The ERG results were also abnormal.

Keywords: Tuberous Sclerosis; Retinal Dystrophy; Electrophysiology; Visual Evoked Potential; Electroretinography.

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Introduction

Tuberous sclerosis complex (TSC) is a disorder affecting the skin, brain, heart, kidneys, lungs, and eyes in ways that can vary across the lifetime¹. Epilepsy is one of the main symptoms among affected individuals with TSC, causing a high rate of morbidity².

A characteristic of TSC patients is drug-resistant epilepsy³. Vigabatrin may significantly reduce seizure frequency in people with drug-resistant focal epilepsy⁴. It is well known that vigabatrin affects the visual system, mainly inducing visual field defects that can be diagnosed using perimetry exam⁵. In addition to perimetry, electroretinography (ERG) and visual evoked potential (VEP) results may be abnormal in patients undergoing vigabatrin treatment⁶.

Here, we report the ERG and VEP examination results of a case of TSC with probable retinal dystrophy and a long history of vigabatrin usage.

Case Report

A twelve-year-old girl with TSC was referred to Basir Eye Clinic, Tehran, Iran, for an electrophysiological examination of her visual system, including VEP and ERG exams. At the age of ten, the patient suffered from severe seizures and was admitted to the hospital. She underwent various anticonvulsive medications without success until the neurologist prescribed vigabatrin, which was successful in controlling the seizures.

In the right eye VEP exam, the latency of the P100 peak was 92 milliseconds, whereas for the left eye it was 100 milliseconds, showing an 8-millisecond difference between the two eyes. Although the VEP P100 peak was within the normal range (the normal value for the VEP P100 peak is about 100 milliseconds in our laboratory), there was a notable discrepancy

between the eyes.

In ERG, the values for the voltage of the b-wave were 61 μ V and 51 μ V for the right and left eyes, respectively (the normal value for b-wave voltage in ERG should be above 100 μ V). Therefore, there were pathological changes in the retinas of both eyes.

The patient underwent perimetry along with VEP and ERG examinations. The perimetry also showed pathological changes.

While reviewing the patient's medical records, the patient's mother informed that their family suffers from retinal dystrophy. The condition was mild in the mother's case. This raised the assumption that the changes in the patient's retina and visual pathway could be either due to the side effects of vigabatrin or hereditary factors from her mother's side.

Discussion

A twelve-year-old girl with TSC was referred for an electrophysiological examination of her visual system, including VEP and ERG exams. She had a long history of using vigabatrin to control her seizures. Vigabatrin, an antiseizure medication used to treat epilepsy and other seizure disorders, can cause several vision-related side effects, including visual field constriction, blurred vision, decreased vision, blue-yellow color blindness, eye pain, and uncontrolled eye movements^{7,8}.

Electrophysiological testing among patients with a history of vigabatrin usage may reveal a normal VEP response, ERG abnormalities, and a reduced Arden Index on EOG testing⁷. We observed similar findings in our patient.

Other antiepileptic drugs have also been reported to negatively impact visual quality among patients. For example, Shushtarian et al.,⁹ reported a case of a 27-year-old female who suffered from sudden total blindness

in both eyes. She had a history of receiving various medications for different illnesses, including epilepsy⁹. She reported the sudden self-termination of levetiracetam, and within one week after the self-withdrawal of the drug, she completely lost vision in both eyes. A complete examination of the patient, including a visual evoked potential test, led to a diagnosis of cortical blindness⁹.

Conclusion

The serious visual side effects of long term antiepileptic drugs usage, although rare, should be considered and patients using these drugs should be routinely examined to rule out these side effects.

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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 manuscript.