Case Report

Visual Evoked Potential Findings in an Eight-Year-Old Girl with Triple-A Syndrome

Seyed Mohammad Masoud Shushtarian *[1], PhD; Ahmad Shojaei [2], MD

1. Department of Biophysics and Biochemistry, Faculty of Advance Science and Technology, Tehran Medical Sciences, Islamic Azad University, Tehran, Iran.
2. Basir Eye Health Research Center, Iran University of Medical Sciences, Tehran, Iran.

***Corresponding Author:** Seyed Mohammad Masoud Shushtarian
E-mail: mshushtarian@yahoo.com

Abstract

Triple A syndrome (Allgrove syndrome) is an autosomal recessive disorder characterized by alacrima, achalasia, and Addison's disease. We report an 8-year-old girl diagnosed with Triple A syndrome and abnormal visual evoked potential findings.

Keywords: Triple A Syndrome; Visual Pathway; Visual Evoked Potential.

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Introduction

Allgrove syndrome, or Triple A syndrome, is a progressive neuro-endocrine syndrome of unknown etiology with an autosomal recessive inheritance pattern [1]. It is characterized by achalasia, alacrima, adrenal insufficiency, and autonomic and neurologic alterations [1]. The estimated prevalence of this syndrome is 1 in 1,000,000 persons [1].

There is little literature on the ophthalmic manifestations of these patients, and it is mostly confined to case reports. Brooks et al.,[2] reported a case of a 12-year-old boy with the classic systemic features of Triple A syndrome and several prominent ophthalmic features, including accommodative spasm, dry eye, superficial punctate keratopathy, and pupillary hypersensitivity to dilute pilocarpine [2].

Visual evoked potential (VEP) findings of Triple A syndrome have been discussed in a case report by Dipinto et al., [3]. They reported the ophthalmic examination of two siblings with Triple A syndrome, including visual evoked response (VER), which showed mild bilateral impairment [3].

In the present case report, we describe the VEP findings in an 8-year-old girl with Triple A syndrome.

Case Report

An eight-year-old girl was referred to Basir Eye Clinic, Tehran, Iran, for VEP recording. The medical history of the patient showed that she was suffering from Triple A syndrome, including achalasia, alacrima, and Addison's disease. Her best corrected visual acuity (BCVA) was 5/10 in the right eye and 2/10 in the left eye. She complained about her vision, which could not be explained as her fundus examination was normal. Nonetheless, she was able to distinguish the fixation point on the center of the monitor necessary to record the pattern reversal checkerboard VEP. A well-formed and reliable pattern was obtained. The VEP peek 100 wave latency times were 115 msec and 125 msec in the right and left eyes, respectively.

Discussion

Allgrove syndrome or Triple A syndrome is characterized by a triad of alacrima, achalasia, and Addison's disease. An eight-year-old girl was referred to Basir Eye Clinic in Tehran, Iran, for a VEP test. Despite having a normal fundus we observed abnormal VEP findings in this girl.

VEP arises from the visual pathway of the visual system, indicating that the patient under study had visual pathway involvement along with Triple A syndrome.

Similar to our findings Moschos et al., [4] have reported three subjects from a Greek family affected by Triple A syndrome. All patients were tested using VEP along with other ophthalmic examinations [4]. It was observed that in two of the subjects, bilateral optic atrophy was present, and VEP results were pathological.

Conclusion

Triple A syndrome is a rare disease which may be accompanied by visual pathway involvement detectable using visual evoked potential testing.

Authors ORCIDs

Seyed Mohammad Masoud Shushtarian:
https://orcid.org/0000-0002-6387-9046

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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.