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Arnold-Chiari malformation – A case report

Arnold-Chiari malformacija – Prikaz slučaja

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Summary

A case of a 14-year-old girl suffering from intense headaches is presented. At the first pediatrician's examination, the patient had regular findings. Prolonged P100 wave latencies were found by testing the visual evoked potentials. After further examination following repeated headaches, Arnold Chiari type I malformation was diagnosed. Arnold Chiari malformation is classified as downward displacement of one or both cerebellar tonsils through foramen magnum. The most common symptom is occipital or suboccipital headaches that usually happens after hard physical effort. Changes in visual evoked potential latencies can be present before other clinical symptoms and can be a useful tool in the diagnostics of headaches in children and adolescents.

Key words: diplopia, cephalgia, Arnold-Chiari malformation, case reports, visual evoked potentials

Sažetak

Prikazan je slučaj djevojčice u dobi od 14 godina koja pati od intenzivnih glavobolja. Bolesnica je na prvom pregledu neuropedijatra imala uredne nalaze. Pretragom vidnih evociranih potencijala nađene su produljene latencije P100 vala. Nakon daljnjih pregleda koji su napravljeni nakon ponovljenih glavobolja, dijagnosticirana joj je malformacija Arnolda Chiarija tipa I. Malformacija Arnolda Chiarija klasificira se pomicanjem jedne ili obje tonzile maloga mozga kroz foramen magnum. Najčešći simptom je okcipitalna ili subokcipitalna glavobolja koja se obično javlja nakon teškog fizičkog napora. Promjene latencija vidnih evociranih potencijala mogu biti prisutne prije ostalih kliničkih simptoma i mogu biti korisno sredstvo u dijagnostici glavobolje u djece i adolescenata.

Ključne riječi: dvoslike, glavobolja, Arnold-Chiari malformacija, prikaz slučaja, vizualni evocirani potencijali

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Introduction

First described by Hans Chiari in the 19th century, the Arnold Chiari malformation is classically defined as the downward displacement of one or both cerebellar tonsils through foramen magnum.[1] The condition is rather common in the general population, usually in young adults, ranging from 0.5% to up to 3.5%. It can be manifested with symptoms ranging from light headache or neck pain to symptoms of acute brainstem compression. 15 to 30% of adult patient with Chiari malformation type I are asymptomatic. Various comorbidities are also common, most frequently syringomyelia which is present in up to 70% of cases.[2,3]

A positive correlation between raised intracranial pressure and prolonged latencies on visual evoked potentials has been proven in hydrocephalic patients and patients with head injuries.[4] Visual evoked potentials could be useful when evaluating the posterior visual pathway and visual cortex. However, the role of visual evoked potentials in Chiari malformation type 1 has yet to be studied.[5] The purpose of this article is to present a case of a 14-year-old female with frequent intensive headaches and

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prolonged latencies on visual evoked potentials that was later diagnosed as Chiari malformation type I.

Case Report

A 14 year old female presented with frequent, intense, occipital headaches, especially after hard physical effort. She was otherwise healthy and was a professional Judo athlete. She was examined by a neuropediatrician and referred to an ophthalmologist.

Upon the first ophthalmic exam, she had visual acuity on both eyes of 20/20. The color vision, tested with Ishihara plates was 38/38 on both eyes. Slit lamp examination was normal, and a relative afferent pupillary defect was absent. Fundus examination showed normal optic disc and retina. Visual evoked potentials performed on monocular stimulation of the samples were recorded in accordance with the International Society for Clinical Electrophysiology of Vision (ISCEV Guidelines). Testing was done with light samples of size 1.0 and 0.3 degrees using TomeyEP-1000 (TOMEY GmbH Am Weichselgarten Erlangen, Germany).

The results showed prolonged latencies on both eyes with normal amplitudes. She was diagnosed as migraines and prescribed pain killers.

Four months after the diagnosis, the patient lost consciousness after her usual Judo training. Her vital signs were normal. Other symptoms included intense headache, vomiting and double vision. She was admitted at the Pediatric Clinic of University Hospital Split. Once more, a complete ophthalmic exam was performed. Visual acuity on both eyes was 20/200. Testing of extra ocular motility revealed bilateral abduction deficit with horizontal binocular double vision. The orthoptic exam showed + 8 prism dioptre esotropia in primary position. In the Hess Lancaster test, she had hypofunction of both lateral recti muscles, with hyperfuncion of medial recti muscles. Visual evoked potentials were repeated, and latencies even more prolonged.

Magnetic resonance imaging described a 7 mm herniation of cerebellar tonsils through the foramen magnum, syringomyelia was not present on the cervical spine MRI. She was diagnosed as Chiari malformation type I. Neurosurgical evaluation dismissed the need for a surgical intervention and she was instructed to rest and pain killers were prescribed. All of her symptoms, including double vision and visual acuity were resolved during the next few days. She was advised to avoid any hard physical effort, including her Judo training. Visual evoked potentials were done for the 3rd time 6 months after the diagnosis (Table 1).

On her routine check-up, one month after hospitalization, she stated that her headaches were less frequent, she had no double vision and complete ophthalmic examination was normal.

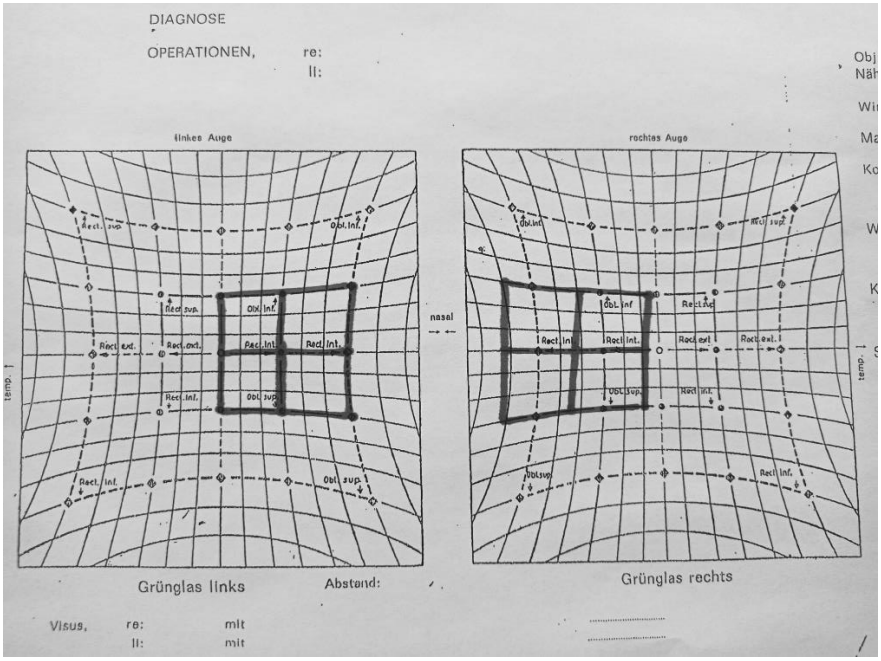

Figure 1 Hess- Lancaster test showing hypofunction of both lateral recti muscles and hyperfunction of both medial recti muscles.

Slika 1. Hess-Lancaster test pokazuje hipofunkciju oba lateralna ravna mišića i hiperfunkciju oba medijalna ravna mišića

Table 1 Results of visual evoked potentials testing using 1 and 0.3 degrees checker.
Tablica 1. Rezultati vidnih evociranih potencijala dobiveni korištenjem podražaja od 1 i 0,3 stupnja

		Right eye *Desno oko*		Left eye *Lijevo oko*	
	Pattern size *Veličina uzorka*	Amplitude *Amplituda* (mV)	Latency *Latencija* (ms)	Amplitude *Amplituda* (mV)	Latency *Latencija* (ms)
1st testing	1º	18.1	112.1	18.6	112.4
	0.3º	17.2	113.4	17.0	113.5
2nd testing	1º	15.5	116.8	15.9	116.2
	0.3º	13.0	115.6	13.1	115.0
3rd testing	1º	18.9	111.1	19.0	111.3
	0.3º	17.2	111.0	111.0	111.0

Discussion

The overall prevalence of headache in children and adolescents is estimated to be around 60%. The most common types are tension headache and migraine.[6] Taking this into consideration, it is very important to diagnose it properly and efficiently. Headaches caused by the Chiari type 1 malformation are usually occipital or suboccipital, short in duration (less than 5 minutes) and are most commonly provoked by cough or other Valsalva- like manoeuvres.[7] Visual evoked potentials, as non-invasive and fast method, could potentially be useful when diagnosing headaches in adolescents. Prolonged latencies, with normal amplitude, are observed in visual evoked potentials, prior to clinical changes in visual acuity in idiopathic intracranial hypertension.[8] There is also a positive correlation of prolonged latencies with elevations of intracranial pressure.[4] It is our opinion that a child and adolescent with a headache and abnormal latencies of visual evoked potentials should undergo further diagnostic examinations, especially MRI. Prolonged latencies of visual evoked potentials must not be disregarded as irrelevant findings.

Double vision in this case was associated with transient bilateral sixth nerve palsy. Miki T et al. in 1999 reported a case of a 12-year-old patient with similar symptoms. Mechanism of bilateral abducens nerve palsy in their case was due to strangulation and downward traction of the pons- oblongata junction.[9] Other cases of transient bilateral sixth nerve palsy in patients with Chiari malformation have also been reported and they usually require operative treatment.

In this case report, the aim is to show the need for visual evoked potentials testing in patients with headaches, which is a non-invasive method, but whose results may suggest the suspicion of serious neurological brain diseases, in this case the Arnold Chiari malformation.

A few authors conducted somatosensory testing (SEP) in patients with Arnold-Chiari's syndrome, but we did not find that the testing visual evoked potentials was performed, so we cannot compare our results with the other ones. SEP (somatosensory evoked potentials) are a neurophysiological technique for examining the sense of touch. They are most often used in the diagnosis and monitoring of diseases and damage to the spinal cord and brain.

Conclusion

Visual evoked potentials could be a safe, fast and non-invasive method in diagnosing headaches in children and adolescents. Prolonged latencies in patients with headache could be a sign of raised intracranial pressure and requires further diagnostics, as was the case with our patient.

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