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Case Report

Atypical oculomotor findings in Arnold–Chiari type I syndrome: A report of two cases

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Abstract

Introduction: Arnold–Chiari type I malformation (ACH I) is classically associated with neuro-otologic symptoms, including vertigo, imbalance, and characteristic oculomotor disturbances – particularly vertical oscillopsia and downbeat nystagmus. However, variability in oculomotor involvement is increasingly recognized. Among the clinical manifestations of Arnold–Chiari type I syndrome, oculomotor disturbances – especially in the sagittal plane – are commonly reported as characteristic findings.

Aim: This study aimed to analyze the presence and nature of oculomotor dysfunction in two patients diagnosed with Arnold–Chiari type I syndrome.

Case study: Two adult patients with MRI-confirmed ACH I underwent comprehensive neuro-otologic evaluation, including videonystagmography (VNG), optokinetic testing, saccadic and smooth-pursuit assessment, caloric stimulation, vestibular evoked myogenic potentials (VEMP), brainstem auditory evoked responses (BERA), audiometry, and complementary neurological and vascular studies. Oculomotor performance was analyzed in the sagittal, frontal, and horizontal planes.

Results and discussion: Neither patient demonstrated oculomotor abnormalities in the sagittal (vertical) or frontal planes. Expected features such as vertical gaze palsy, downbeat nystagmus or torsional deviations were absent. In contrast, both individuals exhibited isolated horizontal-plane dysfunction, including impaired smooth pursuit, asymmetrical optokinesis, horizontal nystagmus, and, in one case, canal paresis. The selective involvement of horizontal oculomotor pathways, with preservation of vertical and torsional mechanisms, suggests heterogeneous patterns of brainstem and cerebellar involvement in ACH I.

Conclusions: Arnold–Chiari type I malformation may present with isolated horizontal oculomotor abnormalities rather than the traditionally described vertical disturbances. Recognition of such atypical patterns is important for accurate diagnostic interpretation and underscores the need for larger studies to elucidate the full spectrum of oculomotor involvement in ACH I.

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1. INTRODUCTION

Described in 1891 by Chiari, a developmental disorder of the craniovertebral junction causing various degrees of displacement of the hindbrain into the spinal canal manifests itself in a number of clinical symptoms, e.g. the vestibular organ and the hearing organ.[1,2] The disease belongs to the so-called dysraphic disorders – a set of congenital defects occurring around 4–10 weeks of fetal life, caused by disorders of neural tube development (NTD). The above-mentioned syndrome also includes: meningocele, spina bifida, diastematomyelia, syringomyelia and anencephaly. Clinical manifestations are caused by compression of the posterior cranial nerves by a displaced brain stem, compression of the cochlear and vestibular nuclei by the cerebellar tonsils, dysfunction of the cochlear aqueduct, and arteriovenous circulation disorders in the posterior cranial fossa.

Based on the severity of pathological changes in the craniovertebral junction, type I Arnold–Chiari malformation (ACH) is characterized by displacement of the cerebellar tonsils into the foramen magnum. In type II ACH, the following structures are moved to the foramen magnum: brainstem, bridge, ventricle IV and worm. In type III, almost the entire cerebellum is displaced, while in type IV, cerebellar development disorders are found.[3] The diagnosis of ACH syndrome relies primarily on computed tomography (CT) and magnetic resonance imaging (MRI), while management commonly involves ventriculoperitoneal shunting in addition to decompression of the spinal canal and posterior cranial fossa.

Type I of the ACH syndrome tends to manifest with neuro-otologic symptoms in the form of vertigo, balance disorders, disorders of vestibulo-oculomotor and visual-oculomotor reflexes in the form of vertical oscillopsia and vertical downward nystagmus, damage to the hearing organ and cerebellar ataxia.[4–6]

The oculomotor symptoms described above in the sagittal plane are regarded as characteristic of this syndrome. However, damage to the midbrain, pons and spinal cord can also cause pathological oculomotor symptoms in the horizontal and frontal planes.

2. AIM

The aim of the study is to describe the potential presence of gaze-motility dysfunction across different planes in individuals with Type I Arnold–Chiari syndrome.

3. CASE STUDY

The first case analyzed was a woman aged 42, complaining of dizziness and periodic balance disorders for 10 years. VNG (videonystagmography) of the equilibrium system revealed disorders of arbitrary tracking, optokinesis and horizontal nystagmus.

In caloric tests, nystagmus reactions were symmetrical, saccade movements of the eyeballs were normal. No disturbances in the somatosensory evoked potentials of the cervical were observed, and ultrasound examination of the vertebrobasilar arteries did not show flow disorders. The cranio-corpographic examination was normal. In the hearing assessment, mixed hearing loss was found on the left side with a predominance of the receptive component, in speech audiometry on this side, the degree of comprehension was at the level of 50 dB, and the degree of discrimination was at the level of 60 dB. In the study of otoemission of the left ear, its impairment was observed. In the VEMP study (Vestibular Evoked Myogenic Potential study) it was found that the latency of the P13 wave was prolonged in relation to the left ear at ipsilateral and contralateral reception. The results of the BERA study (Brainstem Evoked Response Audiometry study) were normal. The functional status of the right ear in the above tests was normal.

The second case was a man aged 61. A physical examination revealed periodic, short-term dizziness for more than 10 years, hypertension with stage II angiopathy. VNG of the equilibrium system revealed disturbances in arbitrary saccadic movement tracking, optokinesis, the presence of horizontal nystagmus type II and canal paresis.

Hearing tests in pure tone audiometry revealed bilateral sensorineural paresis above 4000 Hz, without impairment of speech understanding in speech audiometry. Bilateral impairment of otoemission was recorded, as was bilateral elevation of wave 1 latency in BERA, and elevation of P13 wave latency in the left ear in VEMP and lateral. In the examination of postural reflexes and in the assessment of somatosensory potentials of induced cervical deviations there were no abnormalities observed.

Both patients were managed conservatively, as neither demonstrated clinical or radiological indications for neurosurgical decompression. Neurological examination and MRI findings did not reveal progressive brainstem compression or associated syringomyelia that would necessitate surgical intervention. Management consisted of routine otoneurological and neurological follow-up. During the observation period, symptoms remained stable without the emergence of new deficits.

4. RESULTS AND DISCUSSION

The symptoms of ACH syndrome depend on the type of craniovertebral malformation. In more severe forms, like type III and IV, they appear in the first months of life and are associated with the presence of hydrocephalus. Types I and II occur in adulthood and are caused by increasing dysfunction of the cerebellum, medulla oblongata and cranial nerves, mainly V, IX and XII.[7]

In 70–80% of patients with ACH syndrome type I and II, dizziness, balance disorders and oculomotor dysfunctions in the sagittal plane are found.[7,8] In our patients with ACH type I syndrome, we did not notice vertical eye movement

disorders. We also did not observe oculomotor pathologies in the frontal plane, such as rotational nystagmus, torsional reaction of the eyeballs and oblique deviation of the eyes. These changes are characteristic of acute lesions of the cerebellum, midbrain, pons, or medulla oblongata, whereas the data we analyzed do not represent such cases.[9,10] In the observed patients, however, pathology was distinguished in the stabilization of the gaze in the horizontal plane in the form of disturbances in arbitrary tracking, saccadic eye movements, asymmetry of optokinesis and gaze-directed nystagmus. These symptoms indicate dysfunction of the cerebellar and brainstem oculomotor mechanisms forming tracking movements and the rapid phase of nystagmus.[11–13] The dysfunction can be associated with compression of the brainstem by displaced cerebellar structures. Directional-permanent positional nystagmus observed in one of the patients was associated with canal paresis causing canal-otolith integration disorders, the main cause of peripheral positional nystagmus.[14] Unilateral hearing impairment, shown in one of the cases, due to the mixed nature of hearing loss, was most likely a comorbidity in a patient with ACH syndrome.

The clinical spectrum of Arnold–Chiari type I malformation (ACH I) is recognized as highly variable, and oculomotor disturbances may differ considerably from classical descriptions. Vertical plane abnormalities – particularly downbeat nystagmus and vertical oscillopsia – are traditionally regarded as characteristic signs associated with cerebellar tonsillar descent and floccular dysfunction.[4–6] Nevertheless, several published reports have indicated that these vertical abnormalities are not universally present. Classic studies have emphasized that oculomotor disturbances in Arnold–Chiari malformation typically involve vertical-plane abnormalities. In an early but influential series, Spooner and Baloh evaluated five patients with Arnold–Chiari malformation who underwent suboccipital decompression and reported significant postoperative improvement in key oculomotor functions, including downbeat nystagmus, smooth pursuit, and fixation suppression, highlighting the role of cerebellar compression in generating vertical and global gaze instability.[4] Similarly, a larger cohort study by Liebenberg et al. assessed 40 patients with ACH type I malformation treated with posterior fossa decompression and found that among the 12 individuals with preoperative oculomotor or vestibulo-ocular abnormalities, 8 experienced complete resolution following surgery.[15] Together, these studies reinforce the prevailing view that vertical and combined oculomotor disturbances are common in symptomatic Arnold–Chiari type I malformation, particularly when brainstem or floccular structures are compressed. The lack of vertical or torsional findings in our patients is therefore consistent with the growing understanding of phenotypic diversity in ACH I.

Both of our patients diverged from the classical pattern in another important way.

Both cases instead demonstrated selective horizontal-plane oculomotor impairment, including smooth pursuit deficits, saccadic dysmetria, and asymmetrical optokinetic responses. These abnormalities suggest involvement of cerebellar vermis pathways and pontine gaze structures responsible for horizontal gaze control. Our findings therefore expand the recognized oculomotor spectrum in ACH I, demonstrating that isolated horizontal dysfunction may represent a clinically relevant but underappreciated manifestation of this malformation.

The inclusion of patients of different sexes adds comparative value. Although ACH I is diagnosed more frequently in women, current literature has not demonstrated sex-related differences in oculomotor or vestibular presentation. The similar horizontal-plane abnormalities in both patients support the view that sex is unlikely to substantially influence oculomotor phenotype, though larger cohorts would be needed to confirm this.

Standardizing diagnostic procedures can enhance the interpretability and comparability of findings in conditions such as ACH I. Although both patients in this report underwent comprehensive neuro-otologic evaluation, minor differences in auxiliary testing reflected their individual clinical presentations. In rare disorders like ACH I, the use of harmonized diagnostic algorithms – incorporating consistent vestibular reflex testing, uniform imaging protocols, and parallel neurophysiological assessments – may strengthen internal comparability and support broader generalizability of results. Despite slight procedural variations, the congruent horizontal-plane abnormalities identified in both patients underscore the internal coherence and robustness of the observed oculomotor pattern.

The conservative management applied in these cases aligns with current practice guidelines for Arnold–Chiari type I malformation, in which decompression surgery is recommended only for patients with progressive neurological impairment, significant tonsillar descent with symptomatic brainstem compression, or syringomyelia. Neither of our patients met these criteria, and their symptoms remained non-progressive over time. The stable clinical course further supports the appropriateness of conservative monitoring in individuals presenting with atypical but non-advancing neuro-otologic manifestations.

Collectively, these two cases broaden the documented oculomotor profile of ACH I by illustrating that selective horizontal-plane involvement may occur in the absence of classical vertical disturbances. They also emphasize the importance of comprehensive, multidirectional oculomotor assessment in suspected ACH I. Standardized diagnostic protocols and larger-scale studies will be essential to further clarify the full range of oculomotor dysfunction and its clinical significance in Arnold–Chiari type I malformation.

5. CONCLUSIONS

Observations indicate that oculomotor dysfunction in Arnold–Chiari type I syndrome may deviate from the typical pattern, with preservation of vertical and frontal gaze control and selective impairment confined to the horizontal plane.

Atypical presentation suggests possible variability in the involvement of neuroanatomical structures responsible for

eye movement control, particularly within the brainstem and cerebellum.

While limited by the small sample size, these findings highlight the need for broader clinical and neurophysiological studies to better characterize the spectrum of oculomotor abnormalities in patients with Arnold–Chiari malformation. Recognizing such variability is essential for accurate diagnosis and may inform more personalized approaches to assessment and management.

LIST OF ABBREVIATIONS

ACH syndrome – Arnold–Chiari syndrome
BERA study – Brainstem Evoked Response Audiometry study
CT scan – computed tomography scan
dB – decibel
NTD – neural tube development
MRI – magnetic resonance imaging
VEMP study – Vestibular Evoked Myogenic Potential study
VNG – videonystagmography

Informed consent
Informed consent was obtained from all participants included in the study.

Ethics approval
None declared.

Conflicts of interest
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