Morphomeric Evaluation of Arnold Chiari-Malformations: The Importance of Neurophysiological Diagnosis

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Abstract

Arnold Chiari, also known as Chiari malformation (CM), is a congenital anomaly of the cranio-vertebral junction and hindbrain and is characterized by varying degrees of displacement of posterior pit structures from the foramen magnum to the spinal canal with or without other associated intracranial or extracranial defects such as hydrocephalus, syrinx or spinal. In this malformation group In this process, neuronal dysfunction is observed in the brainstem, cerebellum, and cranial nerves as a result of changes in the spinocerebellar axis due to pathologies such as inferior displacement of the cerebellar vermis, medullary fold formation in the dorsal midbrain, medullary curling, and tectal beaking. Evoked potentials (EPs) are important for clinico-topographic and neurophysiological/neuropathological evaluation. In this review, we aim to review CM neuro-radiologically at the diagnosis stage and to emphasize the importance of neurophysiological recognition of CM in the early stage, especially in asymptomatic patients.

Keywords: Arnold-chiari malformation type • Brain auditory evoked potentials • Cranial MRI • Somatosensory evoked potential • Visual evoked potential

Introduction

CM comprises various pathologies that have in common anatomical deformities of the brainstem and cerebellum. Hans Chiari, an Austrian pathologist, was the first to describe the conditions. His initial manuscript published in 1891, described Chiari malformations 1, 2, and 3. In an 1896 publication, Dr. Chiari postulated the pathogenesis of CM and described Chiari 4 malformation [1-3]. These anomalies are characterized by downward elongation or displacement of the cerebellar tonsils or the vermis into the cervical spinal canal. Abnormalities can be associated with CM, such as hydrocephalus, syringomyelia, spina bifida, hydromyelia, kyphosis, scoliosis, and tethered cord syndrome. In addition, CM may be associated with a number of cranial developmental malformations, known examples of which are craniosynostosis, Dandy Walker, Ehlers-Danlos syndromes and Klippel-Feil syndrome. Classically, the CMs are classified into 4 types: Chiari types 1, 2, 3, and 4. Chiari malformation type 1 (CM1) is by far the most common type of Chiari malformation. CM1 is characterized by extension of the cerebellar tonsils by at least 5 mm below the foramen magnum. Chiari malformation type 2 (CM2) is found in patients with myelomeningocele and involves a greater degree of hindbrain displacement, which may include the cerebellar vermis, brainstem, and fourth ventricle. CM3 is an encephalocele of the posterior fossa with herniation of portions of the cerebellum and brainstem into the encephalocele sac. CM4 is aplasia or hypoplasia of the cerebellum [4]. By Dr. Jerry Oakes and his colleagues, 2 additional types of Chiari malformation have been described; Chiari malformation 0 (CM0) which have syringomyelia without displacement of the cerebellar tonsils, caudal displacement of the cervico-medullary junction and intradural obstruction of cerebrospinal fluid (CSF) flow and Chiari malformation 1.5 is a severe variant of CM1 in which there is a caudal displacement of the brainstem in addition to the cerebellar tonsils below the foramen

magnum [5-7] (Table 1). At the diagnosis stage, demonstrating the tip of the cerebellar tonsils 5 mm below the foramen magnum, associated possible syrinx, and regional malformations on magnetic resonance imaging (MRI) is crucial [8]. Also in CM evaluation, myelography in whic patients that MRI cannot be obtained, CT or x¬rays of the neck and head for preoperative cranocervical structure evaluation and Cine¬MRI for cerebrospinal fluid dynamic evaluation can be used.

Table 1. Summary of types of Chiari malformations.

Description	Chiari Malformation Type
Syrinx without caudal descent of cerebellar tonsils. Syrinx resolves with posterior fossa decompression	0
Caudal displacement of the cerebellar tonsils at least 5 mm below the foramen magnum	1
Caudal displacement of the brainstem and cerebellar tonsils below the foramen magnum	1.5
Caudal displacement of hindbrain structures below the foramen magnum in a patient with myelomeningocele	2
Posterior fossa encephalocele containing brainstem and cerebellar tissue	3
Aplasia or hypoplasia of the cerebellum	4

Evoked potentials are the neuro-sensory response of the nervous system to sensory stimuli using the computer-averaging technique and are distinguished from each other by stimulus modalities: brainstem auditory evoked potentials (BAEP), visual evoked potentials (VEP), and somatosensory evoked potentials (SEP). EPs have been used especially in early stage CM patients as CM-1 as initial diagnostic support and are especially useful for determining the extent of neurologic involvement in asymptomatic or oligo-symptomatic cases. They are also useful for identifying changes that may indicate surgical intervention in the follow-up in whom no symptoms and detected incidentally, as well as oligo-symptomatic patients, is only a few studies, describe the findings of BAEP and SSEP testing in CM-1 [9-12] and unfortunately none for VEP. In this review, a clinico-radiological view of patients with CM will be presented and the importance of using EPs in patients diagnosed with early stage CM and their place in the prognosis will be mentioned.

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Neurological Symptoms and Physical Examination

CM-1 is the mildest of the hindbrain malformations and a number of subgroups have been defined. It is also called primary cerebellar ectopia. The cerebellar tonsils have herniated into the upper cervical spinal canal more than 3 mm from the level of the foramen magnum. This downward slide blocks the circulation of CSF between the spinal cord and the intracranial. In this way, it can cause fluid accumulation in the spinal cord called syringomyelia or hydromyelia. The incidence of the disease in the general population is less than 1 per thousand. In most of the cases, there are no complaints or findings, and it usually occurs by chance as a result of MRI scanning that area for another reason.

This downward slide blocks the circulation of CSF between the spinal cord and the intracranial. In this way, it can cause fluid accumulation in the spinal cord called syringomyelia or hydromyelia. It may occur in 20%-70% of patients with scoliosis and hydromyeli, and it occurs especially in children [13].

CM-2 is a more severe and complex abnormality than CM-1, accompanies spinal dysraphism and is often seen in childhood. The cerebellar tonsils, inferior vermis, and IVth ventricle are displaced caudally from a shallow posterior fossa through a wide foramen magnum, which interrupts the dynamic flow of the CSF and cause hydrocephalus [14]. Tentorium cerebelli and falx cerebri are hypoplastic, with fenestrations and protrusion of the cerebellum from the tentorium opening due to the large incisor. The fourth ventricle appears flattened in the sagittal plane, the lateral recesses have disappeared and displaced caudally, and a tube shaped, elongated, narrow (tube like) fourth ventricle is seen. The choroid plexus may be outside the 4th ventricle (ectopic choroid plexus). Aquaduct stenosis, atresia, bifurcation are common. Lateral enlargement of the ventricles is asymmetrical in many cases. Atria and occipital horns are almost always larger than frontal horns is wide. This condition is known as colpocephaly. Partial agenesis in the corpus callosum, pathologies such as hypoplasia or complete agenesis is seen. Headache due to increased intracranial pressure as a result of hydrocephalus. Opening of sutures and hammered copper appearance on X-ray detected. Scoliosis, cervical spinal canal enlargement, anterior or posterior elements fusion, basilar invagination, has platy base. Tethered spinal cord syndrome, split spinal cord malformation may accompany the picture.

CM-3 is a very rare anomaly. It is defined as the herniation of posterior pit structures such as the cerebellum, brain stem, and 4th ventricle from defects in the lower occipital or higher cervical bone [14]. It has a poor prognosis among others [15].

CM-4 is characterized by the absence or small size of the cerebellum hemispheres, changes in the pons, and cystic enlargement in the 4th ventricle-cisterna magna-basal spaces where cerebrospinal fluid collects. It is rarely seen. Its existence is debatable [14,16].

Diagnosis and Treatment

Diagnosis of anomalies of the craniovertebral junction according to indications includes; clinico-genealogical analysis, MRI of the cranio-vertebral junction, brain and spinal cord in 3-D dimensions also MR-angiography is added if necessary, Computed tomography of the cranio-vertebral to assess the bone structures, X-ray examination of the skull and cervical spine, ofhtalmological examination, otoneurologic examination, ıntrauterine-prenatal echography, Transcranial Doppler sonography, evoked potentials are performed [17-20].

Clinico-radiological evaluation

MRI showing the craniovertebral junction is the first method to be chosen for diagnosis [19-23]. Conventional studies are performed with 1.5 or 3-T MRIwith T1 and T2 sequence. In certain special situations, as in the case of patients with suspected cranio-cervical instability or recent trauma, there may be a benefit of obtaining an upright MRI study with or without flexion and extension of the neck. With MRI, in addition to detecting the characteristic dislocation of the tonsils, we can evaluate the presence or absence of pathology (syringomyelia, hydrocephalus). The extent of tonsillar descent below the foramen magnum, based on Barkovich's study, who stated that patients with 5 mm or more of tonsillar descent were more likely to have a Chiari anomaly [24]. In current studies, it is thought that the measurement of tonsillar descent does not necessarily correlate with the clinical picture. The pointed shape of the cerebellar tonsils indicative of compression, the presence or absence of CSF spaces surrounding the tonsils, and evidence of brainstem compression on axial images at the level of the foramen magnum may be as significant as a measurement of tonsillar descent. CineMRI is useful in the evaluation of cerebrospinal fluid dynamics. May demonstrate blockage of flow at the foramen magnum and distinguish symptomatic ACA types 0 and 1 from asymptomatic cerebellar ectopia, as well as clarify the indications for surgical decompression and predict the outcome of surgical treatment [25,26].

Neurophysiological evaluation

Evoked potentials are noninvasive studies that measure the electrophysiological response of the nervous system to different sensory stimuli. EPs have been used in CM-0, CM-1, and CM-1,5 patients as initial diagnostic support and are especially useful for determining the extent of neurologic involvement in asymptomatic or oligo-symptomatic cases. They are also useful for identifying changes that may indicate surgical intervention in the follow-up of patients, especially in children in whom no symptoms were present and CM-1 was detected incidentally, as well as oligo-symptomatic patients. At the time of writing, however, only a few studies, all with very limited and heterogeneous series of patients, describe the findings of BAEP and SSEP testing in CM-1, and most of them refer to their use during intraoperative neurophysiological monitoring (9-12). As a result, at present, there are still many unknowns regarding the role of EPs in the diagnosis and follow-up of CM-0, CM-1, and CM-1,5; the prognostic value of EPs and their relationship with clinical findings and the severity of malformation has not yet been well established.

Most of the studies have been retrospective studies with a limited number of patients and diverse patient populations and focused on especially SSEPs in patients with syringomyelia [27-31]. Restuccia and Mauguière reported findings consistent with those reported by Anderson et al.; Restuccia and Mauguière defined the most common disturbance as change or absence of cervical potential and increase in N13-N20 [30]. In another study, a high percentage of patients (60%) with CM-1 exhibited EP alterations regardless of their clinical or radiological findings and they concluded that BAEP and SSEP studies play an important role in incidentally detected patients with CM (especially in CM-0, CM-1, and CM-1.5) [32]. They concluded that EPs may help to establish objective evidence of subclinical dysfunctions and neurophysiological studies may help to define subgroups of patients who require further testing and follow-up to personalize strategies for the management of incidental and oligosymptomatic patients.

SSEP

SSEPs, elicited from the upper and lower limbs within 30 ms and 60 ms, respectively, of percutaneous electrical stimulation, are considered resulting from action potentials and synaptic potentials from successive anatomic neural generators within the dorsal-lemniscus thalamocortical sensory system [33]. SSEPs are typically named by their negative or positive polarity at the peak latency, and the time of the peak latency (e.g., N20 is a negative deflection in the EEG waveform, usually peaking at 20 ms poststimulus) as typically observed in the normal population. The actual latency value for a SEP may be different from that implied by the component's name [34]. SEPs evaluate the dorsal column-medial lemniscus pathway, including

the spinal cord and brainstem levels. The lemniscal pathways are typically affected in patients with CM, such as displacement of the cerebellar tonsils, hydrocephalus and syringomyelia. In EPs studies, thermalgesic disturbance was the most common finding, especially in patients with spinal involvement such as syringomyelia, while altered SSEPs were found [31,34,35]. Also, in one study, it is found that the degree of tonsillar herniation was statistically significant in predicting abnormal SSEPs [32]. SSEPs were also abnormal in 30.4% of patients in whom CM had been found incidentally.

VEP

VEP is used to assess the visual conduction pathways through the optic nerves and brain. To measure VEP, visual fields are stimulated, usually with a checkerboard visual stimulus, and the evoked response is recorded using surface recording electrodes over the occipital lobe [36]. The visual perception stimulation method was used to manage cortical visual impairment. Many causes that cause changes in cerebrospinal fluid, such as hydrocephalus, can cause changes in VEP [37]. Hydrocephalus is commonly associated with CMs and it can produce marked changes in the EPs. Several groups of researchers have recorded EPs in hydrocephalic patients in attempts to find a sensitive measure of increased intracranial pressure, of associated pathological changes, or as an indication of the need for neurosurgical intervention [38-41]. Although some of the above studies have found consistent correlations between visual evoked potential (VEP) abnormalities and raised intracranial pressure [42], others have found VEPs to be more useful in only monitoring hydrocephalic patients. Ehle and Sklar found that the 15 infants they studied all had abnormally delayed VEPs, which improved quickly post-shunting [38]. Guthkelch et al. found that increased latencies were present only when the hydrocephalus was accompanied by increased head size (above the 98th percentile); hydrocephalic neonates who were normocephalic usually had normal VEPs [40]. In a later study, they also found that hydrocephalic children had slower maturation of the VEP [39]. They repeated the VEP testing post-shunting in infants <4 months and found only small decreases in latency. Thus, several studies have found VEPs to change with shunting, and therefore it is important in myelomeningocele patients who develop hydrocephalus to record EPs following shunting. In one study, VEPs were recorded in 47 infants with myelomeningocele to determine if the evoked potentials reflected the early neurological status and if they had prognostic value as to the children's neurological outcome. VEPs were abnormal in only 55% of symptomatic infants. Of the infants who did not have symptomatic AC malformation, 69% had normal VEPs. Of the patients with normal VEPs, 63% were normal on follow-up; of the patients with abnormal VEPs, 71% were abnormal on follow-up. It was concluded that the VEPs studied early in the neonatal course do not appear to be sufficiently sensitive to be valuable prognostically in these infants [41]. As we know, there are no studies on the use of VEP in adult patients with CM.

BAEP

The brainstem auditory evoked response (BAER) or potential (BAEP) reflects the electrophysiological activity of many neurons in the brainstem auditory pathway following acoustic stimulation. The BAER is the far-field reflection of sequentially activated neurons at successively higher levels of this pathway and can be used to assess peripheral auditory function, also the functional integrity and development of the brain in general in conditions that affect the brainstem auditory pathway. There is evidence to suggest that BAER waves I and II are generated in the extracranial and intracranial portions of the VIIIeVII nerve, respectively [42]. Subsequent waves IIIeVII are generated in auditory centers at gradually higher levels of the pathway, with partially overlapping contributions to individual waves. Wave III is derived from the cochlear nucleus; wave IV is generated in the superior olivary complex, and wave V e together with the negative potential that follows is generated in the region of the lateral lemniscus and possibly inferior colliculus. Waves VI and VII are likely to originate from the inferior colliculus, although the exact origins remain to be determined. This close relationship

between waveforms and anatomical structures makes it possible to localize accurate conduction defects in the brainstem [42]. BAEPs are extremely useful in the diagnosis and localization of a number of brainstem lesions [43]. Quite frequently, they reveal abnormalities even when CT evidence of disease is lacking or inconclusive. In one study, abnormal responses were obtained in 75% of cases. This figure agrees with the 50%-86% given in other reports in the literature [44,45]. Analysis of symptomatic and asymptomatic cases shows that both groups had prolonged III-V inter-peak latency (IPL). Furthermore, in the symptomatic and 64% of asymptomatic patients, the I-III IPL was normal. In another study, the frequency and degree of severity of abnormalities in the auditory pathways in patients with Chiari malformations type I and II was evaluated in 75 patients (48 children and 27 adults) by means of auditory evoked potential evaluation. Among the 75 patients studied, 27 (36%) disclosed CM-I and 48 (64%) showed CM-II. 53 (71%) of these patients showed some degree of auditory evoked potential abnormalities. Tests were normal in the remaining 22 (29%) patients.

Discussion

They concluded as auditory evoked potential testing is a valuable instrument for the diagnosis and evaluation of brain stem functional abnormalities in patients with CM-1 and CM-2. Determining the extent of these abnormalities is important in terms of prognosis and predicting the handicaps of physical therapy or interventional procedures [46]. In this study, the most frequent abnormality found in BAEPs in classic CM-1 was on a cochlear or auditory peripheral level [47]. In another study, it is found that the degree of tonsillar herniation, and lower cranial nerve dysfunction, had a statistically significant influence in predicting abnormal BAEPs [29]. BAEPs were abnormal in 39.1% of asymptomatic patients on a retrocochlear level. They concluded that the more severe distortion of the brainstem structures induced more BAEP abnormalities and, therefore patients with a higher frequency of abnormal BAEPs.

Conclusion

EPs play an important diagnostic role in asymptomatic/oligosymptomatic patients such as CM-0, CM-1, and CM-1,5 and are also useful in determining prognosis before and after surgery in other types of CMs. MRI is widespread available and crucial for the first diagnosis. In patient follow-up and treatment response, only clinical-radiological follow-up will be insufficient in terms of prognosis and guiding treatment. At this stage, it is important to add neurophysiological evaluation to radiological examinations and to determine the necessity of using each one separately. We believe that the importance of EPs will be better understood with large-scale studies in this area.

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Conflicts of Interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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