Progress of Science: Theory and Practice
Volume 1, Issue 1, December, 2024

CLINICAL AND NEUROLOGICAL MANIFESTATIONS OF ARNOLD-CHIARI MALFORMATION

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The review is devoted to the problem of Chiari malformation. The issues of clinical and neurological disorders, surgical treatment of the disease are considered. Clinical polymorphism of MC suggests a detailed study of the disease symptoms and the need for differential diagnosis. The complexity of early diagnostics of MC is due to the similarity of clinical and neurological manifestations of the anomaly with other neurological diseases. Particular attention is paid to modern methods of instrumental examination of the disease, their comparison with each other with a description of the advantages and disadvantages.

Key words: Chiari malformation, magnetic resonance imaging, craniovertebral region, ectopia cerebellum.

According to the literature, respiratory disorders, central apnea, syncope, cough syncope are observed in 4-20% of patients with CM [1, 2]. There are numerous descriptions of isolated cases of nocturnal apnea in adult patients with CM, detected using the polysomnography method [3, 4, 5]. It is assumed that fainting occurs due to a sharp change in intracranial pressure, which can occur after coughing, sneezing, straining, physical exertion. According to studies, the presence of syncope in patients with CM is due to a vascular factor associated with transient compression of brain tissue and vessels of the vertebral-basilar basin at the level of the craniovertebral region. This is confirmed by the cessation of fainting after successful decompression of the posterior cranial fossa [6, 7].

Clinical polymorphism of CM requires a detailed study of the disease symptoms and the need for differential diagnosis. The difficulty of early diagnostics of CM is due to the similarity of the clinical and neurological manifestations of the anomaly with other neurological diseases. Clinical signs of CM in some cases are similar to multiple sclerosis, consequences of traumatic brain injury, vertebrobasilar insufficiency syndrome of vertebrogenic genesis, degenerative diseases of the nervous system, spinocerebellar degenerations, isolated tumors of the posterior cranial fossa and craniovertebral region [8, 9]. In patients with CM in combination with syringomyelia, the first symptoms of the disease are usually secondary to syringomyelia [10].

Thus, the clinical and neurological manifestations of CM are varied and are caused both by herniations of the cerebellar tonsils and by disturbances in the cerebrospinal fluid circulation in the area of the cisterna magna. Variants of cerebrospinal fluid circulation in patients with CM are represented by retro-, supra-, latero-, intracerebellar cerebrospinal fluid volumes and syringomyelic cavities due to the difficulty of passage of cerebrospinal fluid of various natures at the level of the posterior cranial fossa [11, 12].

A certain amount of help can be provided by a system of clinical syndromes formed by a number of authors on the basis of a variety of complaints and objective neurological symptoms, including the most common disorders: hypertensive-hydrocephalic syndrome, suboccipital pain, segmental dissociated sensory disturbances, pyramidal, cerebellar and bulbar disturbances, cranial nerve lesions, brainstem disturbances, scoliosis and dysraphic signs [13]. Most often, a combination of these syndromes occurs [14].

Research methods

Preoperative verification of CM is an extremely important problem of today's surgery. In the diagnosis of craniovertebral region anomalies, in particular CM, radiation methods play a major role. The use of magnetic resonance imaging (MRI) in clinical practice has significantly improved the diagnosis of pathological changes in the brain, including the craniovertebral region. Due to the ability to display the anatomical structures of the brain in three planes and provide contrast of soft tissues without the use of contrast agents, MRI allows us to clarify the nature of the disease and justify the possibility of its successful treatment. MRI has significantly improved the diagnostic capabilities of CM due to the visualization of the craniocervical junction zone on sagittal tomograms and is currently the method of choice [15, 16, 17]. The use of MRI has revealed a wide range of cerebellar tonsil prolapse in patients with CM. Diagnosis of MC in patients with a large malformation does not cause difficulties, but in cases where the position of the cerebellar tonsils is at a borderline level, diagnosis is greatly complicated. A number of authors have conducted studies to measure the descent of the cerebellar tonsils using MRI, which revealed

the norm, relative to which the comparison was carried out, which also made it possible to differentiate MC from concomitant pathology - hydrocephalus, syringomyelia and syringobulbia and other malformations of brain structures [18, 19].

The introduction of the method of phase-contrast magnetic resonance liquorography with reference to the cardiac cycle made it possible to study the movement of liquor in the subarachnoid spaces of the posterior cranial fossa, spinal cord and in the cavity of syringomyelic cysts [20, 21]. These studies made it possible to clarify the pathogenesis

of syringomyelia in patients with CM, as well as the technique of surgical treatment of this disease.

A promising type of MRI is magnetic resonance angiography (MRA), which allows observing blood flow in extra- and intracranial arteries of the brain [22, 23] and assessing collateral blood flow through the circle of Willis.

In the last two decades, the leading positions in diagnostics of lesions of the vascular system of the head have been occupied by ultrasound diagnostic methods: ultrasound Dopplerography (USDG), color duplex scanning (CDS) [25, 26]. Non-invasiveness and high information content of these methods allow them to be successfully used to study the state of cerebral hemodynamics in patients of any age with vascular diseases of the brain, regardless of their condition. An increase in the blood flow velocity in the internal carotid artery (ICA) was noted in patients with vertebral artery syndrome with a significant bilateral decrease in blood flow in the vertebral artery [27]. Increased blood flow in the ICA was also detected on the side of the posterior trifurcation of the ICA [10]. Transcranial Dopplerography (TCDG) described characteristic changes in cerebral hemodynamics in intracranial hypertension - "difficult perfusion" syndrome [28]. Duplex scanning is used in diagnostics of anomalies and variants of the structure of the cerebral arteries [29]. The methods of TCDG and USDG made it possible to differentiate various variants of the structure of the circle of Willis, as well as to identify circulatory disorders in the arteries of the vertebral-basilar basin in patients with bone anomalies of the craniovertebral region [30].

Existing methods for studying the vascular system of the brain, along with undeniable advantages, also have

a number of disadvantages. Thus, the disadvantages of the MRA method include a fairly long period of time required to obtain images (12-17 min), the high cost of equipment and its operation, special requirements for the premises in which the devices are located (shielding from radio frequency interference). It is difficult to use the MRI method to examine patients with psychomotor agitation, claustrophobia, artificial pacemakers, large metal implants, etc. [31].

However, the main and predominant method for studying the craniovertebral region is currently MRI as the most informative method of neuroimaging, facilitating the choice of tactics for managing a patient with CM.

Surgical treatment of Chiari malformation

The main method of treating MC is surgical intervention, since drug therapy using dehydrating, vascular and symptomatic drugs does not have a significant and lasting effect, and after its cessation, the main manifestations of the disease continue to

increase. Surgery is carried out primarily to eliminate the factors blocking the cerebrospinal fluid pathways and restore physiological cerebral and spinal cerebrospinal fluid circulation. All patients with MC in the presence of neurological symptoms should be recommended early surgical treatment immediately after diagnosis. The risk group for MC should include patients with a combination of cervical-

occipital localization, vestibular, bulbar and pyramidal disorders. Indications for surgical treatment are the following factors: addition of hydrocephalic changes in hypertension syndrome and prolapse of the cerebellar tonsils according to MRI data and the formation of severe dropsy; ineffectiveness of previously conducted conservative treatment, deterioration in the patient's quality of life caused by increased cephalgia during minor physical exertion and psychoemotional stress, an increase in cerebrospinal fluid dynamics disorders according to MRI data; cerebellar dysfunctions; accumulation of cerebrospinal fluid under the tentorium, the formation of pathological cerebrospinal fluid volumes in the pontocerebellar cistern, inside the cerebellar hemisphere, behind the cerebellum according to

CT, MRI. Movement disorders in MC are usually detected in patients with the presence of a syringomyelic cavity, which can be considered as one of the main mechanisms for the formation of conduction disorders; damage to the cranial nerves in MC with a combination of pathological accumulation of cerebrospinal fluid volumes in various parts of the brainstem [32, 33, 34, 35].

Currently, various modifications of the Gardner operation are performed, including resection trepanation of the posterior cranial fossa, dissection of the dura mater and, in some cases, the arachnoid membrane with or without resection of the cerebellar tonsils, with or without reconstruction of the large occipital cistern. [36]. In addition, cerebrospinal fluid shunting operations are additionally performed as one of the stages of surgical intervention. In the presence of syringomyelia, in some cases, dissection and drainage of syringomyelic cysts is performed. With the introduction of microsurgical equipment, it has become possible to perform coagulation and, in some cases, resection of the tonsils [37]. In the case of a combination of MC and syringomyelia, surgical intervention is aimed at eliminating the neurological deficit caused by the presence of the syringomyelic cavity. American neurosurgeons suggest performing ventriculodrainage surgery as the main method of treating combined CM with hydrocephalus. This treatment option has been reflected in a number of other studies [38, 39]. Another approach to brainstem decompression is to perform plastic surgery of the cisterna magna by suturing a dura mater transplant. In addition, experimental work was conducted with the introduction of kaolin into the cisterna magna and the development of a cicatricial adhesive process in this area. The need to

restore cerebrospinal fluid circulation at the level of the cisterna magna is also confirmed by the introduction of dyes into the cisterna magna and the transposition of contrast into the perivascular and perineural spaces of the upper cervical roots [40].

Despite the variety of surgical treatment options, the operation cannot be fully planned, since many intraoperative findings cannot be verified before the operation. In particular, the adhesion process around the tonsils, the posterior inferior cerebellar artery, and the fixation of the upper cervical spinal cord require additional surgical manipulations [41]. Rare findings may include

changes located at the bottom of the fourth ventricle (glial or arachnoid cysts, glial or choroidal nodes, subpendymomas), optic nerve edema, when reconstructive surgery can be supplemented with fenestration of the optic nerve sheaths [42] can be attributed.

According to various authors, the effectiveness of surgical treatment of CM is 70-90% [43, 44, 45]. Thus, in the work carried out by the staff of the Department of Neurology of the Belarusian State Medical University on groups of patients depending on the disorder of cerebrospinal fluid dynamics, high effectiveness of surgical treatment was shown (cerebrospinal fluid shunting, reconstructive operations on the posterior cranial fossa, as well as combined operations were used). In the group of patients with an isolated form of CM, 92% of patients experienced complete or partial regression of neurological symptoms, while in most cases reconstructive operations with resection of the cerebellar tonsils were performed (50%). In the group with a combination of CM with syringomyelia, the main operation was a reconstructive operation with resection of the tonsils (89%), and the percentage of recovery and improvement in condition was 61.1% and 38%, respectively, while complete or partial regression of neurological symptoms occurred in all patients of the study group. In the group with additional cerebrospinal fluid volumes in the posterior cranial fossa, a favorable outcome of surgical intervention was observed in all patients [46, 47].

In the postoperative period, there is a regression of radicular, conductive and segmental disorders, as well as a decrease in the size of the syringomyelic cyst. In patients with occlusive hydrocephalus, a decrease in the size of the cerebral ventricles and early regression in the postoperative period of hypertensive-hydrocephalic syndrome are noted. However, there are known cases of various complications after surgery. The most dangerous of them is edema of the brain and upper cervical spinal cord after reconstructive operations on the posterior cranial fossa and after cerebrospinal fluid drainage operations [48, 49], which occurs most often in the early postoperative period. A cicatricial-adhesive process can also form after decompression of the brainstem with subsequent occlusion of cerebrospinal fluid pathways and progression of hydrocephalus [50].

Thus, surgical intervention is an adequate method of treatment and monitoring of the patient's condition.

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