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CHIARI MALFORMATION I TYPE COMPLICATED WITH SYRINGOMIELIA - CURRENT STATE OF THE PROBLEM (LITERATURE REVIEW)

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Abstract:

The article presents a review of the literature on the current understanding and topical issues of type I Chiari malformation complicated by syringomyelia. Particular attention is paid to the etiology, epidemiology, mechanism of development, types of diagnostic methods and currently existing methods of treatment, their advantages and disadvantages. The paper also presents various theoretical options for the chronological stages of the development of syringomyelia in Chiari type I malformation and methods for eliminating this pathology.

Keywords: Chiari malformation, syringomyelia, craniovertebral junction, foramen magnum, osteodural decompression

INTRODUCTION

Chiari malformation type I (MK I), both an independent nosology and complicated by syringomyelia, is one of the complex and problematic anomalies of the craniovertebral junction and is characterized by an imbalance between the nerve structures and the total volume of the posterior cranial fossa. The manifestation of clinical manifestations is associated with the descent of the tonsils of the cerebellum and other structures of the cerebral table, depending on the type of malformation, below the conditional line of the foramen magnum (BSO). One of the most common complications of type I MK is syringomyelia (SM). Syringomyelia is a chronic process morphologically manifested by the formation of fluid-filled cavities in the spinal canal [1, 2, 3, 4, 5, 25].

The incidence of MK varies among the population of the CIS countries and, according to different authors, ranges from 3.3% to 8.2% per 100,000 populations. In approximately 1% of the healthy population, an MRI examination of the brain reveals a descent of the cerebellar tonsils 5 mm below the BZO line. This indicator is sufficient to make a diagnosis of type I MK, according to modern criteria for this pathology [25, 26, 27, 28]. Globally, type MK I affects about 0.01–0.04% of the population. The ratio of male and female in total is almost the same, but one of the features of the epidemiology of this pathology is the predominance of males among the inhabitants of the northeast and the predominant number of females in the countries of the southwest. This originality explains the ethnic features of the MK I type. Spinal syringomyelia occurs in 80-92% of patients with MK I [1, 2].

In modern literature, there is more and more information that the disease is more likely acquired than congenital, as many experts previously believed, but due to the lack of research, this assumption cannot give an unambiguous answer in favor of one or another hypothesis.

The history of the study of the issue began in the 19th century, when J. Clenland in 1883 noticed a pathological elongation of the structures of the brain stem and the descent of the cerebellar tonsils into the spinal canal in 9 deceased infants [6, 7]. Later, in 1891, the German pathologist Hans von Chiari described a clinical case, which was characterized by the descent of the cerebellar tonsils and hernial protrusion of the tonsils, as well as the brain stem below the BZO line. In 1894, J. Arnold also found a similar change in the craniovertebral region in patients [1, 8]. But there are later data, already in 1593-1674. Tulpa described a herniated hindbrain characteristic of myelomeningocele

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[1]. If all the cases described above occurred in the children's contingent, then in 1938 Connell and Parker for the first time revealed the descent of the cerebellar tonsils below the BZO line and, as a result, developed hydrocephalus in adults. They associated this condition with CSF flow blockage by ectopic cerebellar tonsils as a "plug" in the foramen magnum [5, 7].

ETIOLOGY AND PATHOGENESIS

Currently, there are two main theories of the origin of MK I type: heredity or genetic conditioning and an acquired mechanism, starting from the early postnatal period (late prenatal, intranatal and postnatal periods). Despite new data in favor of the acquired origin of MK, the theory of the genetic condition of pathology still remains a more reliable option among specialists and scientists.

G. Chiari explained the occurrence of this pathology by hydrocephalus, i.e. hydrodynamic disturbances in combination with insufficient growth and volume of the skull bones [7, 8]

With the improvement of research methods in the field of medicine, there has been a closer look at the etiology and mechanisms of development of KS, as well as syringomyelia. J. Cleland believed that the occurrence of MC is associated with dysgenesis of the nervous structures and bones of the skull. Barry A. and J. Cleland studied five human embryos and found that brain outgrowth shifts the cerebellum caudally leading to MS [5, 7, 8, 9]

Subsequently, Nyland H. and Krogness K.G. After studying five adult patients, they concluded that the relatively rapid growth of the cerebral hemispheres and the relatively slow growth of bones, especially the occipital, lead to a pronounced decrease in the volume of the posterior cranial fossa and the descent of the main structures in the BZO [16, 17]

Another amazing theory was put forward by Kuether T.A. According to his theory, vitamin D-resistant rickets causes disproportionate growth of the skeletal system of the skull and suggested that this condition can lead to Chiari type I and type II anomalies [12]

Finally, the isolation of the PAX I, PAX 9 genes contributed to the development of a more detailed genetic theory. It is these parts of the chromosomes that contain the information code responsible for the growth and formation of the structures of the craniocervical region [21]. This theory was also confirmed by the studies of Stovner L.J. three monozygotic twin sisters, their mother and three of their daughters, who had KS, and one of the twins also had syringomyelia [22].

Hida K. and a number of other researchers suggest that birth trauma is one of the main etiological factors in the development of MC. After numerous observations Valiulin M.A. and Larionov S.N. developed the concept of bone-neural disproportion, which is based on the fact that CM develops as a result of birth trauma [11, 25, 28].

In 2005, a new theory developed by Roth M., Royo-Salvador M.V. appeared on the world stage of neurology and neurosurgery. – in their opinion, the development of scoliosis, syringomyelia, and Chiari malformation is associated with the "hard terminal filament" (Fixed terminal filament), which leads to overstretching of the spinal cord and, accordingly, to the appearance of symptoms of the above conditions [19, 20]

Gardner W.J. and his hydrodynamic theory of the occurrence of syringomyelia - gave a clear statement of the question of the formation of syringomyelitic cysts of the spinal cord in MC. According to his theory, syringomyelia develops as a result of a constant systolic pulse impulse created by the vascular plexuses of the ventricular system, which prevents the fusion of the central spinal canal and gradually leads to local expansion [9, 10].

The dissociation of craniospinal pressure proposed by Williams B. also explains the progression of symptoms of MC and syringomyelia. The essence of this theory is the difference in pressure in the intracranial and extracranial spaces, after conducting Valsalvi tests, the author showed that when sneezing, coughing and straining, high intrathoracic pressure is transmitted to the epidural veins of the spinal cord and a pressure wave is formed that directs approximately 8 ml of CSF into the subarachnoid space of the cranial cavity , and the obturated BSO pushes the cerebrospinal fluid into the open central spinal canal and, accordingly, the resulting pressure wave leads to the formation and expansion of syringomyelitic cysts of the spinal cord [23, 24]

Based on the data presented, it can be concluded that syringomyelia can be both communicating and non-communicating in CM. The abundance of theories about the etiology of KS and syringomyelia allows us to conclude that the consensus and the final answer to the questions posed have not yet been developed [27].

CLINIC

The manifestation of clinical symptoms begins on average from 15 to 45 years. There are only a few cases when symptoms appeared at a younger or senile age [24, 25, 26]. The nonspecificity of the onset of the disease is the main reason for the omission of early diagnosis and the development of complications.

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The first signs of MK I type are headache, which is a sign of more than 70% of neurological diseases and, accordingly, this circumstance significantly complicates timely diagnosis. The later stages of this pathology are characterized by symptoms such as dizziness, impaired motor skills, sensory, swallowing, speech and autonomic disorders.

Headache with localization in the cervical-occipital region and spread throughout the head is the first cause in patients seeking medical help. According to different authors, approximately 81-100% of patients with type II CM complicated by syringomyelia suffer from cervicocranialgia [1, 2, 20, 24]. A characteristic feature of cranioservicoalgia is the initiation of Valsalvi headaches in similar situations such as sneezing, straining, bending over, yawning, laughing, and straining. At the same time, the Valsalvi maneuvers themselves can easily cause short-term or long-term headache [23, 24].

Among the nonspecific clinical manifestations of MK I, one of the most frequent and severe are coordination disorders that occur at different stages of the disease and have polymorphic manifestations [28]. Coordination disorders in VM I are characterized by a wide range of cerebellar-stem disorders in combination - compression and liquorodynamic genesis. Among the subjective disorders of coordination in MK I, the presence of non-systemic dizziness is considered a classic [25]. Coordination disorders are the most common symptom in patients with MK I, along with headaches, reaching 57.9% during routine neurological examination and increasing to 84.1% with additional coordination tests. the most sensitive technique is provocative tests for dizziness. [23, 25, 28].

Cerebellar symptoms are manifested by static and dynamic ataxia, impaired coordination of movements, horizontal nystagmus, chanted speech, deliberate trembling, hypermetry, megalography, asynergy, and decreased muscle tone [1, 25].

Among the symptoms of MK I type, including both early and late, bulbar disorders can be highly informative: swallowing disorder, visual impairment, nystagmus (Ocular phenomena include retroorbital pain, photomorphopsia, photophobia, blurred vision, diplopia and limited visual fields). tinnitus, speech disorders, nighttime snoring, sleep apnea, etc. [14, 16].

Syringomyelitic symptoms: atrophy of individual muscles (often the distal shoulder and biceps, in some cases accompanied by a tear of its tendon) occurs in 55.6% of patients. Dissociated sensory disorders are observed in 50-91% of patients and are usually localized in the cervicothoracic segments in the form of a "jacket" and "half jacket". Trophic disorders of the skin of the hands in the form of hyperkeratosis, cracks, poorly healing wounds, osteoarthropathy of the destructive-hypertrophic type - in 23.1% [14, 15, 19, 20] sensitivity disorders: in the form of paresthesia, hyperesthesia, pain, analgesia or anesthesia, dysesthesia, reduced temperature sensitivity. Complaints of pain manifestations with localization in the arm, cervical-occipital region or chest occur in 90% of patients. The type of pain disorder is most often aching in nature and has a different degree of severity. Complaints of paresthesia in the form of numbness, sensations of cold, chilliness of the body area are observed in approximately 39% of patients. Well-being Loss of temperature sensitivity can often be associated with the presence of painless burns [13, 14, 26].

Perhaps the most striking semiotic sign during the initial examination of patients with CM is the dysraphic status: low hairline in the occipital region, angioma in the occipital region, symptom of a "short neck", "high" palate, asymmetry of the chest, "sandal-shaped". "gap" between the first and second toes, Friedreich's foot deformity, ear deformity (very massive lobes, "satirical ear", fused lobes, poorly differentiated auricle, etc.), microgenia, accessory papillae, cervical ribs, progenia [9, 13, 24, 25]

In addition to the above symptoms of damage, such signs as memory impairment, decreased attention, anxiety, nervousness, depression, depression of various types can play an important role in the diagnosis [17, 18].

METHODS

Before the invention and introduction of magnetic resonance imaging into the clinic, the detection of KS and syringomyelia was one of the most difficult tasks for neurosurgeons. Therefore, today the main method for diagnosing MC is MRI examination of the brain and spinal cord. Even an inexperienced surgeon, after looking at the results of an MRI of a patient with this pathology, can make a preliminary diagnosis of Chiari malformation, but, having the skills to decipher the data obtained, the volume and size of the PCF, the specialist doctor is able to accurately determine the type, stages and specific treatment tactics.

Morphometric angular and linear measurements in CM make it possible to accurately determine the dimensions of the PCA and adjacent structures (Fig. 1), [24, 25]

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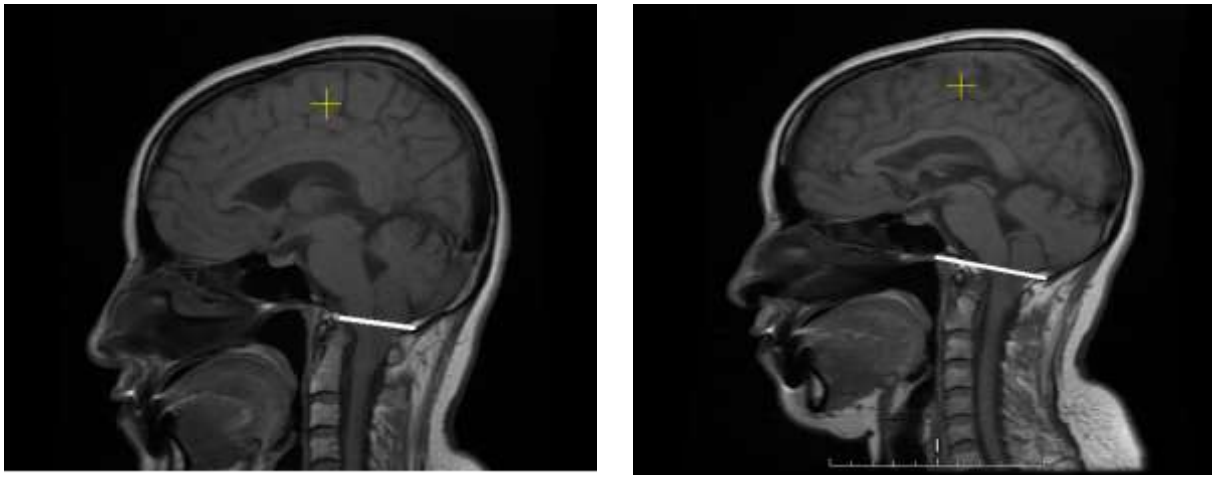

Fig. 1. Imaginary lines of McRae and Chamberlain.

One of the preferred methods for diagnosing patients with CM and syringomyelia is phase-contrast MRI synchronized with the cardiac cycle, which helps to determine the patency of subarachnoid CSF spaces [13, 25, 26]. It was with the help of this method that the connection between syringomyelia and MC was established.

A promising type of MRI is magnetic resonance angiography (MRA), which makes it possible to observe blood flow in the extra- and intracranial arteries of the brain and evaluate collateral blood flow using the circle of Willis system [28].

MRI, like other research methods, is not without contraindications. Among them are claustrophobia, metal implants, psychosis, etc. [17, 22, 28]

In recent years, more and more attention has been paid to electroneurophysiological methods in MC and syringomyelia. Multimodal evoked potentials are informative for detecting conduction disturbances in the structures of the brainstem and upper structures of the spinal cord [1, 2, 25]. The use of these studies before the operating and postoperative periods makes it possible to evaluate the effectiveness of the therapy performed, as well as the volume of the planned operation.

TREATMENT

Currently, the main method of treatment of MK I type and syringomyelia is surgical. Specific types of operations depend on the form, type and associated anomalies. Perhaps the most common surgical method today is osteo-dural decompression of the craniovertebral region. But in each case, a certain tactic must be applied. The list of surgical methods, along with classical bone-dural decompression, includes: bone decompression, bone decompression with dissection of the outer layer of the dura mater, bone-dural decompression in combination with syringotomy, syringo-bypass surgery, endoscopic triventriculostomy and ventriculo-shunt surgery [1, 2, 3, 4, 10, 11, 17, 26, 27].

But still, studying the world literature, one can find more specific features of operational approaches for this pathology. Listed below are a few reliable and validated methods. Sometimes, despite a successful operation, regression of syringomyelia may not be observed, in such cases it is necessary to perform bypass surgery [20, 22, 26, 28].

Filling the obex (valve) with a piece of muscle can be used in case of a confirmed form of syringomyelia due to MK, i.e. liquorodynamic disorders. A striking example can be found in the work of Gardner. The essence of his operations consisted of a wide craniectomy of the posterior fossa, opening of the IV ventricle, sealing the obex with a piece of muscle and leaving the dura open. The author reported 5 deaths after 74 operations [9, 10].

In some cases, during the operation, the infringement of the tonsils of the cerebellum is detected and in such cases a subpial resection of the tonsils of the cerebellum must be performed. It is also possible to plan a subpial resection before the operation period, when the descent reaches C2 [25, 26].

The material for dura plastic surgery deserves special attention. Today, for this purpose, various types of plastics are used: artificial material, autofabric and others. The outcome is dilemmatic. According to some authors, when using artificial DM, in the postoperative period, signs of aseptic meningitis (up to 35%) and pseudomeningocele (1%) often occur. But in the presence of adhesions and arachnopathy, the best option is an artificial material [7, 13, 19].

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CONCLUSIONS

Thus, at present, the most adequate treatment method for type I MK complicated by syringomyelia is the surgical correction of the craniovertebral junction. In 90% of cases, after surgery, the condition of patients improves markedly and there is a regression of clinical and neurological symptoms. Nevertheless, there are many unresolved issues regarding the choice of treatment tactics, which requires a more thorough study of the issue and improvement of the results of treatment of patients with this pathology.

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