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CLINICAL CASE REPORT

A RARE CLINICAL CASE OF SYRINGOMYELIA PROGRESSION IN THE PRESENCE OF CHIARI I MALFORMATION FOLLOWING THE SURGERY

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ABSTRACT

Today, an Arnold-Chiari malformation is defined as a developmental pathology of the craniovertebral junction manifested by a discrepancy between the volume and contents of the posterior cranial fossa, which in turn leads to compression of neurological structures and changes in the cerebrospinal fluid circulation. There are several theories of the correlation between Chiari malformation and syringomyelia, but the exact mechanism of syringomyelia development remains unclear.

This article describes a clinical case of treatment of a child with Chiari I malformation and syringomyelia within the cervical and thoracic segments of the spinal cord; after complete posterior fossa decompression, syringomyelia progressed in the early postoperative period with the development of a severe neurological deficiency. Since there is no standard treatment of such postoperative complications, a decision was made on the expectant management of the patient. From the twentieth day of the postoperative period, the patient showed complete regression of the neurological deficiency and positive MRI dynamics of syringomyelia.

The presented clinical case raises such issues as not only the pathophysiology of syringomyelia progression after complete posterior fossa decompression, but also the determination of patient management tactics in case of a complicated postoperative course of the disease.

The presented clinical case is of interest due to the rarely described aggravation of syringomyelia with enhancing neurological symptoms in the early postoperative period after complete posterior fossa decompression.

Our observation suggests that the expectant management of the patient, despite syringomyelia progression with neurological deficiency aggravation after posterior fossa decompression, allowed a favorable long-term outcome of Chiari I malformation.

Keywords: syringomyelia progression, Chiari I malformation, craniovertebral junction anomaly, posterior fossa decompression

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КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ

РЕДКИЙ КЛИНИЧЕСКИЙ СЛУЧАЙ ПРОГРЕССИРОВАНИЯ СИРИНГОМИЕЛИИ НА ФОНЕ АНОМАЛИИ КИАРИ I ТИПА ПОСЛЕ ОПЕРАТИВНОГО ВМЕШАТЕЛЬСТВА

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РЕЗЮМЕ

На сегодняшний день мальформация Арнольда-Киари определяется как патология развития краниовертебрального перехода, проявляющаяся несоответствием объема и содержимого задней черепной ямки, что в свою очередь приводит к компрессии неврологических структур и изменению характера ликворотока. Существует несколько теорий корреляции между мальформацией Киари и сирингомиелией, однако точный механизм развития сирингомиелии остается предметом для дискуссии.

В статье описан клинический случай лечения ребенка с аномалией Киари I типа и сирингомиелией шейных и грудных сегментов спинного мозга, у которого после проведенной декомпрессии задней черепной ямки «полного объема» в раннем послеоперационном периоде возникло прогрессирование сирингомиелии с развитием грубого неврологического дефицита. Учитывая отсутствие стандартных подходов в лечении таких послеоперационных осложнений, принято решение о выжидательной тактике ведения пациента. С двадцатых суток послеоперационного периода у пациента отмечен полный регресс неврологического дефицита и положительная магнитно-резонансная томография (МРТ) динамика сирингомиелии.

Представленный клинический случай поднимает не только вопросы патофизиологии прогрессирования сирингомиелии после выполнения декомпрессии задней черепной ямки «полного объема», но и определение тактики ведения пациента в случае осложнённого послеоперационного течения заболевания.

Интерес представленного клинического случая заключается в том, что описано редко встречающееся усугубление сирингомиелии с нарастанием неврологической симптоматики в раннем послеоперационном периоде после выполнения декомпрессии задней черепной ямки «полного объема».

Основываясь на данных нашего наблюдения, выжидательная тактика ведения пациента, несмотря на прогрессирование сирингомиелии с усугублением неврологического дефицита, после декомпрессии задней черепной ямки позволила добиться благополучного отдаленного результата течения заболевания, связанного с аномалией Киари I типа.

Ключевые слова: прогрессирование сирингомиелии, аномалия Киари I, аномалия краниовертебрального перехода, декомпрессия задней черепной ямки

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Соблюдение этических стандартов: в работе соблюдались этические принципы, предъявляемые Хельсинкской декларацией Всемирной медицинской ассоциации (World Medical Association Declaration of Helsinki, 1964, ред. 2013). Информированное согласие получено от всех участников исследования.

Финансирование: финансирование данной работы не проводилось.

Конфликт интересов: все авторы заявляют об отсутствии явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

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INTRODUCTION

Currently, Arnold-Chiari malformation is defined as a pathology of the development of the craniovertebral junction, manifested by a mismatch in the volume of the posterior cranial fossa and brain structures, which in turn leads to prolapse of the cerebellar tonsils below the Chamberlain line, compression of neurological structures within the cranio-vertebral junction and impaired cerebrospinal fluid dynamics of varying severity [1].

In clinical practice, the most common is type I Chiari anomaly. According to the literature, it occurs with a frequency of 1 per 1000 newborns, with a female prevalence of 1.3 to 1 [2]. In 20–85 % of cases, Chiari I is potentially associated with syringomyelia, more often affecting the cervical region, followed by a combined lesion of the cervical-thoracic region (Fig. 1.). However, there is currently no consensus explaining the etiology and progression of syringomyelia.

In the neurological aspect, Chiari type I anomaly remains asymptomatic in most cases, which creates certain difficulties in diagnosing the disease and is detected when performing magnetic resonance imaging (MRI) for other reasons. When the disease manifests, the most common symptom in both adults and children is headache and segmental violations of temperature sensitivity are much less common [3].

According to modern concepts, MRI of the central nervous system is the main and widely used diagnostic method that allows to obtain an image of the anatomy of the craniovertebral junction with the detection of hydrocephalus and/or syringomyelia, as well as fixed spinal cord syndrome [4].

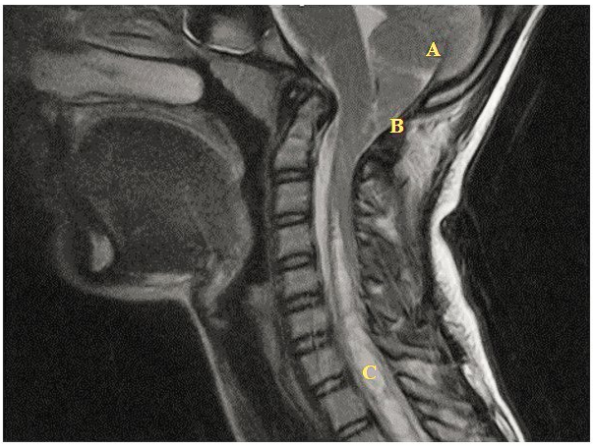

Fig. 1. Arnold-Chiari anomaly type I: A – cerebellum; B – prolapse of the cerebellar tonsils below the foramen magnum; C – syringomyelia at the cervical level.

The main method of treatment of patients with craniovertebral junction anomaly is surgical intervention, which is indicated by the presence of neurological symptoms associated with both syringomyelia and the insertion of the tonsils of the cerebellum [5]. Surgical intervention is aimed at restoring cerebrospinal fluid dynamics at the level of the craniovertebral junction. Optimal access consists in performing a suboccipital craniectomy with decompressive expansion of the foramen magnum, often with laminectomy of the posterior arch C1, and if necessary, the arch C2 of the vertebra, plastic surgery of the dura mater is performed.

Thus, the intervention, designated as decompression of the posterior cranial fossa of the "full volume", is the most common neurosurgical approach aimed at restoring the flow of cerebrospinal fluid at the level of the foramen magnum.

However, there are many questions here: what amount of decompression is required to successfully change the pathology of the cerebrospinal fluid flow, what treatment results are considered satisfactory, what tactics to choose with the further progression of syringomyelia in the postoperative period, what mechanism is responsible for the ineffectiveness of treatment, what is the need and priority of additional surgical interventions [6]?

The clinical example we have given shows the difficulty of determining tactics with an extremely rare aggravation of syringomyelia with the development of a gross neurological deficit after performing a "full-volume" surgical intervention.

Description of the clinical case

Patient T., 17 years old, was admitted to the clinical diagnostic department of the National Medical Research Centre for Oncology with complaints of severe headache in the occipital region. Pain syndrome according to the visual-analog pain scale is 7 points. In the neurological status: cerebral syndrome; tendon and periosteal reflexes in the extremities symmetrical; muscle strength 5 points; segmental sensory disorders were not detected. According to MRI of the central nervous system (Fig. 2): omission of the tonsils of the cerebellum

by 18 mm below the occipital foramen magnum (Fig. 2A). There are no data for hydrocephalus. Syringomyelia of the cervical and thoracic spinal cord with expansion of the central spinal canal up to 6 mm (Fig. 2 B, C). Lack of fixation of the spinal cord at the lumbar level.

The patient underwent surgical intervention in the volume of resection of the posterior edge of the foramen magnum and the posterior semicircle of the atlas (Fig. 3); Y-shaped opening of a significantly hypertrophied dura mater and its subsequent plasty using an artificial dura mater plate from Medtronic. The arachnoid meninges were not opened in connection with the performed tasks of surgical intervention, in order to avoid further formation of the adhesive process.

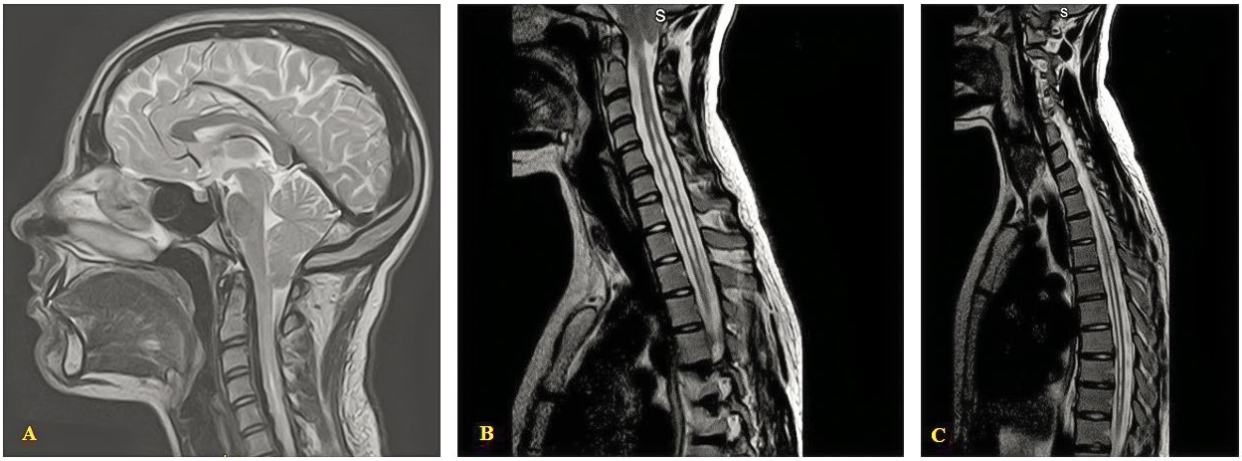

Fig. 2. Data of preoperative MRI of the central nervous system (T2 mode) of the A – brain, B – cervical spinal cord, C – thoracic spinal cord.

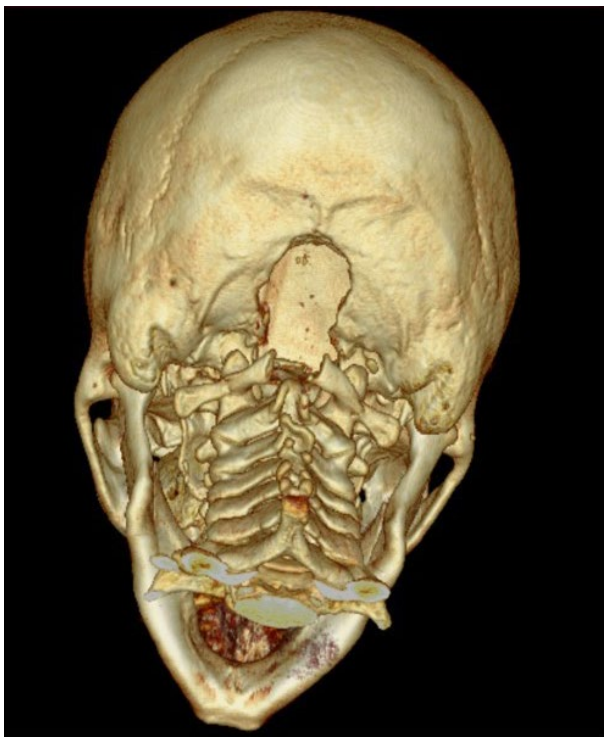

Fig. 3. 3D model of bone decompression reconstruction based on the results of postoperative CT scan of the brain.

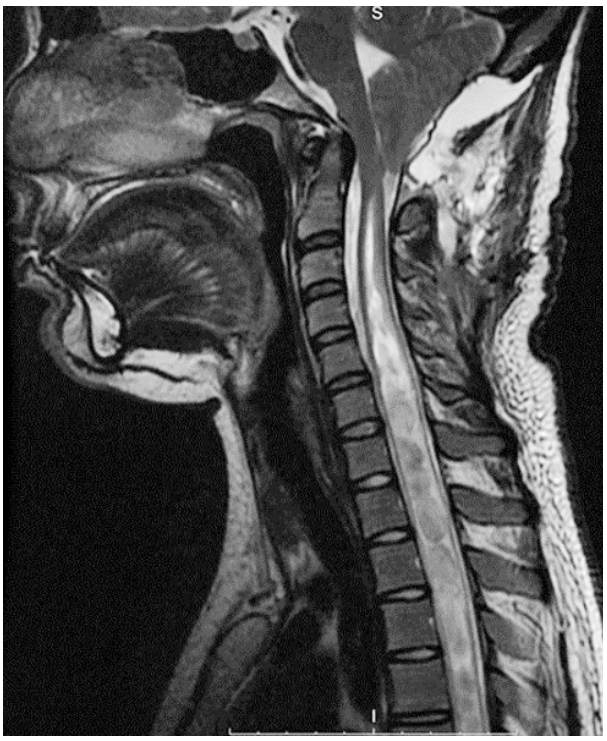

Fig. 4. MRI data (T2 mode) of the brain, cervical spinal cord – 3 days after surgery.

In the early postoperative period, the patient had an increase in cerebral symptoms, the appearance of pronounced muscle weakness in the upper extremities up to 2 points and pronounced hypesthesia in the hands. A control MRI of the central nervous system was performed on the 3rd day after the operation. To exclude the potentially reversible state of "presyrinx", MRI was performed using T1 and T2-weighted sequences [7]. MRI data of the brain and cervical spinal cord: no hydrocephalus; prolapse of the cerebellar tonsils into the surgically expanded occipital foramen magnum remains 18 mm below the large occipital foramen; negative dynamics of syringomyelia of the cervical spinal cord with the expansion of the central spinal canal to 14 mm (versus 6 mm before surgery) (Fig. 4).

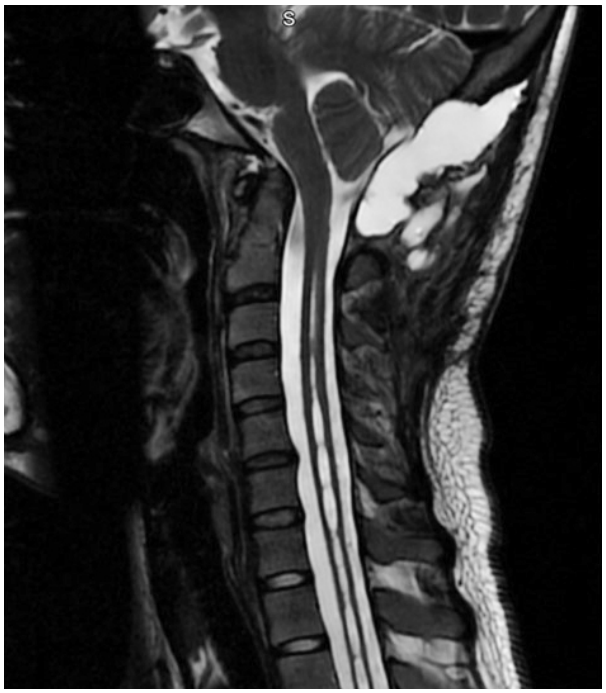

Fig. 5. MRI data (T2 mode) of the brain, cervical spinal cord performed 23 days after surgery.

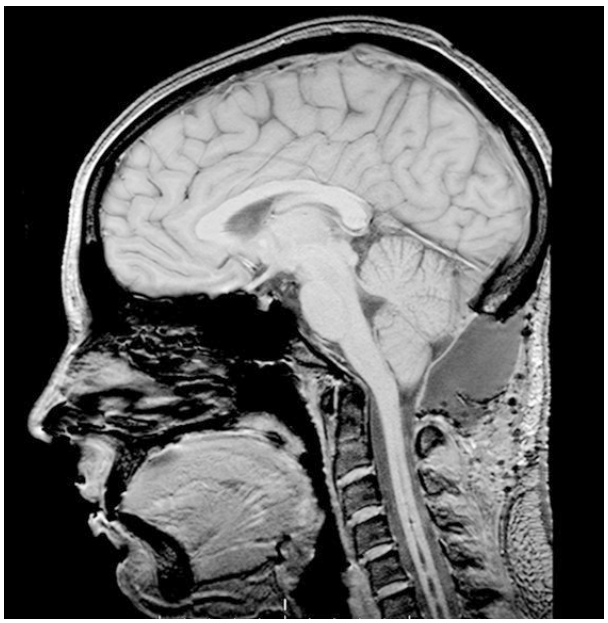

Fig. 6. MRI data (T1 mode) of the brain, cervical spinal cord performed 3 months after surgery.

Given the lack of standard treatment tactics, in this case, a decision was made on conservative management of the patient. Acetazolamide is prescribed in a daily dose of 250 mg. Since the twentieth day after the operation, a positive dynamics of the neurological status was noted in the form of an increase in muscle strength to 5 points, a complete regression of the cerebral syndrome and sensitive disorders.

MRI data of the brain and cervical spinal cord performed on the 23rd day after surgery revealed positive dynamics of the disease: omission of the tonsils of the cerebellum by 11 mm (vs. 18 mm on the 3rd day after surgery); syringomyelia of the cervical spinal cord from the cerebrospinal junction and caudal with the expansion of the central spinal canal to 3 mm (vs. 14 mm on the 3rd day after surgery) (Fig. 5).

Clinically, in the late postoperative period, there is a complete regression of the cerebral syndrome and the absence of focal manifestations.

On the control MRI 3 months after the operation, further positive dynamics was noted: lowering of the tonsils of the cerebellum by 8 mm (versus 11 mm on the 23rd day after surgery); syringomyelia of the cervical spinal cord with expansion of the central spinal canal to 2 mm (versus 3 mm on the 23rd day after surgery) (Fig. 6).

DISCUSSION

Suboccipital craniectomy, resection of the posterior semicircle of the atlas with subsequent plasty of the dura mater without opening the arachnoid medulla is a highly effective method of treating Chiari type I anomaly associated with syringomyelia. However, this method does not exclude a complicated course of the disease with further progression of syringomyelia and aggravation of neurological deficit, which makes it difficult for the neurosurgeon to choose the patient's treatment tactics.

In search of an answer to the question about the possible mechanisms of the progression of syringomyelia after performing a decompressive operation of "full volume", it is worth referring to the work of Aboulker J. (1979), who suggested that the displacement of the cerebellar tonsils downwards causes stenosis of the subarachnoid space at the level of the foramen magnum with the possible formation of syringomyelia. After decompression of the posterior cranial fossa, the tonsils are further displaced downwards due to the weakening of the "support" of the cerebellum [6]. This is also confirmed by the theory of Oldfield E. H. et al. (1994) on the pulsating aggravation of the dislocation of the cerebellar tonsils during each respiratory cycle and cardiac systole, followed by the development of dynamic blockade of the cerebrospinal fluid pathways in the region of the foramen magnum and aggravation of syringomyelia [8-10]. A number of authors, investigating this issue, identified as the basis of the persistent course of the disease: syringomyelia in this group of patients does not always occur due to obstruction of the cerebrospinal tract in the area of the foramen magnum, or the operation did not eliminate obstruction of the cerebrospinal tract in the area of the foramen magnum, which initiates the pathophysiological mechanism of progression of syringomyelia [11].

Based on our observation data, the patient's wait-and-see management tactics, despite the progression of syringomyelia with an aggravation of neurological deficit after decompression of the posterior cranial fossa, made it possible to achieve a safe long-term outcome of the course of the disease associated with type I Chiari anomaly.

CONCLUSION

The given clinical case demonstrates that patients after neurosurgical treatment for type I Chiari anomaly associated with syringomyelia should undergo thorough clinical and instrumental dynamic monitoring for potential progression of syringomyelia. In case of an increase in syringomyelia and aggravation of neurological symptoms in the postoperative period, conservative management of the patient is permissible.

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Contribution of the authors:

Rostorguev E. E. – designed research framework, interpreted the results, edited the text of the paper;

Kuznetsova N. S. – designed research framework, analysed the data, drafted the manuscript;

Maslov A. A. – edited the text of the paper, analysed the results;

Hatyushin V. E. – reviewed the publications on the topic of the article, analysed the data;

Matevosyan B. V. – processing and analysis of the results;

Reznik G. A. – reviewed the publications on the topic of the article;

Pandova O. V. – reviewed the publications on the topic of the article;

Shalashnaya E. V. – edited the text of the paper.