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Original investigation

Investigation of The Improvement Rate Regarding The Herniation of Cerebellar Tonsils Following Shunting Procedures in Patients with Chiari Malformation and Associated Hydrocephalus

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ABSTRACT

Introduction and Aim: Chiari malformation is a congenital anomaly formed as the result of the herniation of posterior fossa structures through the foramen magnum toward the spinal canal. In this study, we aimed to present the improvement rate regarding the herniation in cerebellar tonsils following shunting procedures in patients with Chiari malformation and associated hydrocephalus.

Method: We measured in mm the improvement rate regarding the herniation in cerebellar tonsils in postoperative cervical MRI images obtained following ventriculoperitoneal shunting procedures for hydrocephalus in adult patients with symptoms of Chiari malformation such as balance disorder, dizziness, numbness and loss of muscular strength at the hands who were diagnosed with Chiari malformation and hydrocephalus following cranial and cervical magnetic resonance imaging (MRI).

Results: A total of fifteen adult patients in whom the cerebellar tonsillar herniation was over 5 mm and accompanied by hydrocephalus were included in our study. The measure of herniation in the patients involved in the study was between 1.11 cm and 8 mm. The amount of improvement in herniation following the shunting procedure was between 7.76 mm and 2.2 mm.

Conclusion: In patients with Chiari malformation associated with hydrocephalus, cerebellar tonsillar herniation can regress to 5 mm, which is considered as physiological, and the symptoms related to Chiari malformation can disappear. When Chiari malformation is associated with hydrocephalus, the tonsillar herniation may improve, and symptoms may disappear following the ventriculoperitoneal shunting procedure for hydrocephalus without necessitating posterior decompression for Chiari-related herniation.

Keywords: chiari malformation, tonsillary herniation, ventriculoperitoneal shunt

INTRODUCTION

Chiari malformation, defined as the caudal displacement of structures located in the posterior fossa through the foramen magnum toward the cervical canal, was first described by Cleland in 1883 (1). Subsequently, an Austrian pathologist named Hans von Chiari published his first case series in 1891 and 1896, classifying the malformation according to the extent of herniation; accordingly, four classical types of Chiari malformation were described (2, 3). Later, considering the differences in treatment, definitions such as Chiari malformations Type 0 (Chiari-like malformation), Chiari Type 1.5, Chiari Type V, and Complex Chiari have been added (4–7). The gold standard for diagnosing the Chiari malformation is magnetic resonance imaging (MRI). Radiologically, Chiari malformation is described as the 5 mm or more downward displacement of the cerebellar tonsils through the foramen magnum (8).

Even though the symptoms and signs of Chiari malformation may be related to disorders of cerebrospinal fluid circulation, it has been stated that they might be associated with intracranial lesions as well as hereditary connective tissue disorders. However, there is neither a definite cause nor a relevant theory on which a consensus has been reached yet (9, 10). Hydrodynamics, excessive growth, tension and traction, neuroschisis, developmental arrest, small posterior fossa, and primary mesodermal insufficiency have all been encountered among the theories. However, no single common pathogenetic cause describing all types of Chiari malformation is currently present (11–18).

Starting with the first report of Chiari in 1891, an association of hydrocephalus with Chiari malformation has been reported repeatedly. Some authors have suggested that the circulation of cerebrospinal fluid is obstructed at the foramen magnum,

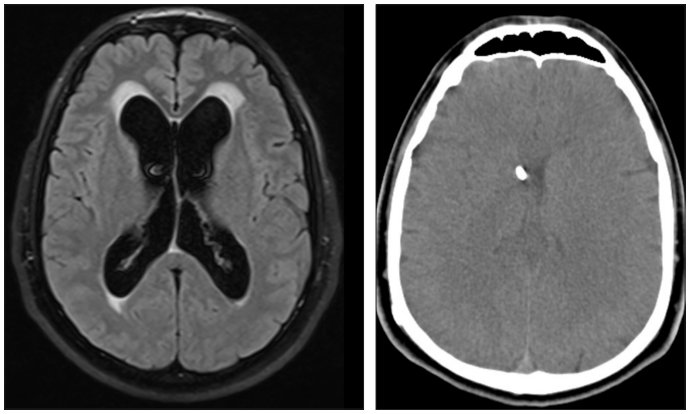

Figure 1. a, b. The preoperative cranial MRI shows the enlargement of the ventricles and edema related to CSF passing to the periventricular regions in T1 axial plane (a). The postoperative cranial MRI shows the reduction of the ventricular dimensions following the ventriculoperitoneal shunting procedure (b).

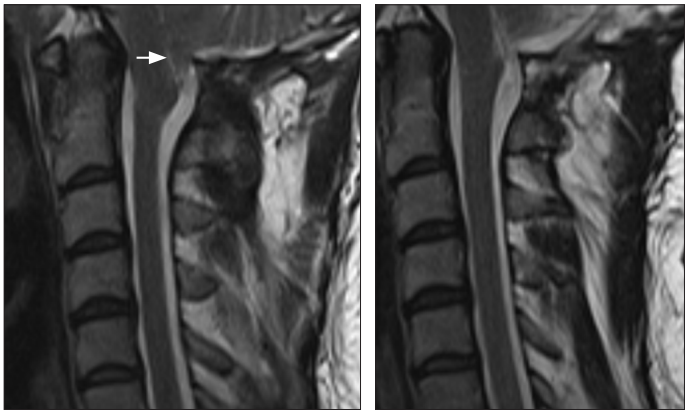

Figure 2. a, b. The preoperative cervical MRI shows in sagittal T2 plane the 1.11 cm-long herniation of the cerebellar tonsils through the foramen magnum towards the spinal canal (a). The postoperative cervical MRI shows in sagittal T2 plane that the 1.11 cm-long herniation of the cerebellar tonsils through the foramen magnum toward the spinal canal improved following the ventriculoperitoneal shunting procedure performed for hydrocephalus (b).

leading to secondary hydrocephalus (19–21), whereas others including Hans Chiari himself have proposed that herniation of the cerebellar tonsils develops secondarily as the result of supratentorial compression due to long-lasting presence of hydrocephalus (2, 22, 23).

The controversial issues related to the pathophysiological mechanisms regarding the Chiari malformation – hydrocephalus association put aside, recently conducted studies have reached a consensus on the subject that, since the treatment of hydrocephalus gets rid of the supratentorial compression, it improves tonsillar herniation, and therefore, symptomatic hydrocephalus, either primary or secondary, should be handled prior to the management of Chiari malformation (8, 24, 25). In this study, we aimed to investigate the improvement rate regarding the herniation of cerebellar tonsils following shunting procedures in patients with Chiari malformation and associated hydrocephalus.

MATERIAL and METHOD

Fifteen adult patients having symptoms such as balance disorder, dizziness, headache, paresthesia at the hands, amnesia, and loss of muscular strength, who had been diagnosed with Chiari malformation and hydrocephalus following cranial and cervical magnetic resonance imaging between 2010 and 2019 were included in the study. The amount of caudal herniation of the cerebellar tonsils during the preoperative period and following the shunting procedure performed for treatment of hydrocephalus were measured and recorded.

RESULTS

The mean age of the patient group included in the study was 38.5 years, ranging from 25 years to 58 years. Nine patients were female, and six were male. In all patients included in the study, the amount of tonsillar herniation was over 5 mm, ranging from 8 mm to 11.1 mm.

All patients had hydrocephalus. A ventriculoperitoneal shunting procedure for hydrocephalus was primarily performed only without decompression of the posterior fossa in all patients. Radiologically, ventricles were determined to be diminished in size after shunting. Figures 1a and 1b show an example regarding the beneficiary effect of ventriculoperitoneal shunting procedure on ventricular dimensions and periventricular edema.

All patients showed a reduction in the amount of tonsillar herniation following the shunting procedure for hydrocephalus. The reduction ranged from 2.2 mm to 7.76 mm. In twelve of fifteen patients who had undergone ventriculoperitoneal shunting, the amount of herniation of cerebellar tonsils decreased to less than 5 mm, which is considered as the physiological value. Figures 2a-2b exhibit example of the beneficiary effect of shunting, showing the MR images of two cases obtained in the preoperative and post-shunting periods. In the remaining three patients, the amount of tonsillar herniation did not decrease to less than 5 mm.

In thirteen patients, improvement of the symptoms was observed (86.7%). On the other hand, in two of the three patients in whom the amount of herniation had not decreased to less than 5 mm, even though some improvement was observed, complete recovery did not occur, and after that, decompression of the posterior fossa was performed. The demographic characteristics, the preoperative symptoms, signs, and the amount of herniation, the postoperative course of clinical features, herniation, and the requirement for decompressive surgery were shown in Table 1.

Table 1. The demographic characteristics, the preoperative symptoms, signs, and the amount of herniation, the postoperative course of clinical features, herniation, and the requirement for decompressive surgery of the patients

Age (years)	Gender	Preoperative clinical features	Preoperative tonsillar herniation (mm)	Post-shunting course of clinical features	Post-shunting tonsillar herniation (mm)	The requirement for posterior fossa decompression
25	F	Headache, dizziness, imbalance	11.1	Decreased	3.3	-
40	F	Headache	8.0	Decreased	3.5	-
33	M	Headache, dizziness, hypoesthesia	9.6	Decreased	3.2	-
56	M	Headache, dizziness, hypoesthesia	10.3	Decreased	4.0	-
32	M	Headache	8.8	Decreased	2.5	-
49	F	Headache	9.2	Decreased	2.8	-
44	M	Headache, hypoesthesia	9.8	Unchanged	6.8	+
27	F	Headache	10.7	Decreased	2.0	-
35	F	Headache, dizziness, hypoesthesia	9.2	Slightly decreased	6.0	-
51	F	Headache	10.3	Decreased	3.9	-
36	M	Headache	8.6	Decreased	4.0	-
40	M	Headache, imbalance	10.4	Unchanged	8.2	+
52	F	Headache, imbalance	10.1	Decreased	3.8	-
58	F	Headache	9.7	Decreased	3.1	-
38	F	Headache	9.8	Decreased	3.6	-

DISCUSSION

Hydrocephalus may accompany the Chiari malformation. Among patients having Chiari I malformation, approximately 15% to 20% have hydrocephalus also (26). We determined in this study that, following the ventriculoperitoneal shunting procedure, the herniation of the cerebellar tonsils reverted to physiological limits and the symptoms improved significantly in most of the patients with hydrocephalus accompanying the Chiari malformation.

The relationship between hydrocephalus and the Chiari malformation has not been completely understood. Regarding the pathophysiology of their association, controversial points of view are currently present. While some of the authors suggest that the interruption of the cerebrospinal fluid circulation at the foramen magnum causes hydrocephalus, others propose that herniation of the cerebellar tonsils develops as the result of supratentorial compression due to long-lasting hydrocephalus (8, 19–21, 24).

Even though the pathophysiology has not reached a consensus, the treatment of hydrocephalus has been commonly considered to have a priority when associated with the Chiari malformation, because, due to its curative effect on supratentorial compression, the shunting procedure leads to an improvement in the severity of tonsillar herniation as determined radiologically in various studies (8, 24). The results of our study were consistent with the literature, demonstrating well in our case series the significant reduction in the size of tonsillar herniation and ventricular dimensions (Figures 1a-b, 2a-b).

A full spectrum of clinical symptoms such as headache, balance disorder, hypoesthesia, and loss of muscular strength may be present in patients with the Chiari malformation. In our patients, all kinds of symptoms except the loss of muscular strength were determined to be present preoperatively. These symptoms were found to improve following the shunting procedure in most of our patients. This finding of ours was consistent with the literature, even though the literature related to the management of hydrocephalus associated with the Chiari malformation has recently focused on the endoscopic third ventriculostomy technique (27), advocating it as an alternative method to ventriculoperitoneal shunting. Various authors such as Massimi et al. and Wu et al. have published their good results with ventriculostomy, reporting that the symptoms, which had been present preoperatively improved significantly during the post-ventriculostomy period (27, 28).

On the other hand, patients in whom herniation of the cerebellar tonsils does not revert to physiological limit following ventriculoperitoneal shunting procedure, headache and gait disturbance might not sufficiently improve, necessitating decompression of the posterior fossa for the Chiari malformation. As the result of our study on our case series involving 15 patients with the Chiari malformation associated with hydrocephalus, we determined that while symptoms of 13 patients had improved following the ventriculoperitoneal shunting procedure, two patients had required to undergo decompression of the posterior fossa due to the Chiari malformation. When these patients were analyzed individually, it was found that the amount of herniation correlated with persistence of symptoms; in both patients, the amount of post-shunting cerebellar tonsillar herniation was above the physiological cut-off value of 5 mm (6.8 mm and 8.2 mm). Surprisingly, in another patient of ours, the amount of herniation decreased from 9.2 mm to 6.0 mm; however, this patient did not require decompression of the posterior fossa, since her symptoms decreased slightly, not being intolerable (Table 1).

As a conclusion, the results of our study supported the hypothesis that in Chiari malformation, the herniation of cerebellar tonsils

develops due to supratentorial compression caused by the long-term presence of hydrocephalus. Second, our study supported the consensus on management of hydrocephalus in Chiari malformation, advocating either ventriculoperitoneal shunting or third ventriculostomy procedures initially in symptomatic cases with Chiari malformation. Multi-center, prospective studies should be conducted for the establishment of standardized algorithms regarding such a complex malformation with a complex pathophysiology.

Informed Consent: Consent was obtained from patients who had surgery to use their results in scientific studies

Compliance with Ethical Standards: Written permission was received from the Ethics Committee of Tekirdağ Namık Kemal University (Approval number:26/12/2019).

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Author Contributions: Concept - MOY, SK; Design - MOY, SK; Supervision - MOY, SK; Fundings - MOY, SK; Materials - MOY, SK; Data Collection and/or Processing - MOY, SK; Analysis and/or Interpretation - MOY, SK; Literature Search - MOY, SK; Writing Manuscript - MOY, SK; Critical Review - MOY, SK

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