Annals of Case Reports
Marc G, et al. Ann Case Rep: 7: 1039
www.doi.org/10.29011/2574-7754.101039
www.gavinpublishers.com

Case Report

Chiari Type I Malformation Associated With Hydrocephalus and Syringomyelia Treated By Endoscopic Ventriculocisternostomy

Gbazi Marc[1], Capel Cyrille[1,2*], Lantonkpode Romaric[1], Carlier Pauline[1], Balédent Olivier[2,3], Peltier Johann[1]

[1]Department of Neurosurgery, Hospital University Centre of Amiens-Picardie, 80054 Amiens, France

[2]CHIMERE UR 7516, Jules Verne University, 80000, Amiens, France

[3]Image Processing department, Hospital University Centre, 80054, Amiens, France

[*]**Corresponding author**: Cyrille Capel, Department of neurosurgery, Hospital University Centre of Amiens-Picardie, Rond Point du Professor Christian Cabrol, 80054 AMIENS, France

Citation: Marc G, Cyrille C, Romaric L, Pauline C, Olivier B, et al. (2022) Chiari Type I Malformation Associated With Hydrocephalus and Syringomyelia Treated By Endoscopic Ventriculocisternostomy. Ann Case Report. 7: 1039. DOI: 10.29011/2574-7754.101039

Received Date: 12 November 2022; **Accepted Date**: 15 November 2022; **Published Date**: 17 November 2022

Abstract

Introduction: The treatment of the Arnold Chiari malformation associated with hydrocephalus is debated, as is the pathophysiology. Phase contrast magnetic resonance imaging (PCMRI) allows non-invasive analysis of cerebrospinal fluid (CSF) and arterial and venous blood flow. This technique can be an aid in the understanding of physiopathological mechanisms and therapeutic decisions. **Clinical Case**: A 41-year-old female patient presented with headaches associated with a tetrapyramidal syndrome, motor disorders, and vigilance disorders. The radiological workup was in favour of a triventricular hydrocephalus associated with a Chiari type I malformation (MCI) and a cervical syringomyelic cavity. An endoscopic ventriculocisternostomy was urgently indicated. Postoperative radiological follow-up showed regression of the ventricular cavities, complete regression of the ptosis of the cerebellar tonsils, and disappearance of the syringomyelia. Phase contrast MRI performed remotely showed aqueducal stenosis and hydrodynamic restitution of the CSF at the foramen magnum. **Conclusion**: The performance of an ETV presents low complication rates with important clinical effectiveness in Chiari malformations associated with hydrocephalus and syringomyelia.

Keywords: Endoscopic Ventriculocisternostomy; Chiari type I malformation (MCI); Syringomyelia

Introduction

Chiari I malformation (MCI) is defined as a descent of the cerebellar tonsils more than 5 mm below the foramen magnum [1, 2]. The clinical presentations are multiple. An association with hydrocephalus is noted in 9% of cases. The association with syringomyelia seems to be more frequent (65%) [3].

The pathophysiology of the association of MCI with hydrocephalus is debated in the literature [4, 5]. Currently, only a few clear data points allow us to understand this context.

Analyses are based essentially on anatomical and preoperative imaging data. Similarly, therapeutic approaches are debated [4-7], probably due to the lack of a clear pathophysiological understanding. An analysis of the dynamics of cerebrospinal fluid (CSF) could allow a different understanding of the pathophysiology of intracranial fluids flows in order to better adapt the therapeutic management.

In the physiological state, the CSF circulates in a pulsatile manner in the sub-arachnoid spaces at the cardiac rhythm. This pulsatility of CSF is achieved by a flush to the perimedullary

subarachnoid spaces in the systolic phase and an inflow of CSF to the intracranial subarachnoid spaces in the diastolic phase. These inflow-outflow phenomena compensate for variations in intracranial vascular volume during the cardiac cycle [8, 9]. This phenomenon is called mobile compliance by several authors [4, 9]. Obstruction of the foramen magnum limits the passive circulation of the CSF towards the spinal subarachnoid spaces, thus a loss of part of the cerebral autoregulation of the intracranial pressure (ICP). A Phase contrast magnetic resonance imaging (PCMRI) study enables us to quantify CSF and blood flow at the craniospinal level [8, 10]. This allows characterization of craniospinal hemodynamics and hydrodynamics. Therefore, the analysis of the pathophysiology of MCI by PCMRI seems relevant.

We report a case of MCI associated with hydrocephalus and syringomyelia that rapidly resolved after endoscopic ventriculocisternostomy (ETV).

Clinical Case

A 41-year-old female patient was referred to the emergency department and was then hospitalised in the neurosurgery department of the Amiens-Picardie University Hospital for vigilance disorders. She presented with headaches that had been progressing for several months. An unusual ronchopathy had appeared at the same time as the headaches. For several days, she had been experiencing walking and balance disorders associated with a tetrapyramidal syndrome and multi-modal sensitivity disorders. Oculomotor analysis revealed bilateral VI involvement. The clinical examination revealed sensory disorders of the right hemisphere without a motor deficit. She rapidly showed the onset of slight vigilance disorders (Glasgow coma scale at 12).

The MRI (Figure 1A) showed triventricular dilatation and effacement of the cortical sulci. It showed a near- complete disappearance of all the cisterns of the infratentorial stage and a descent of the cerebellar tonsils of about 13 mm below the plane of the foramen magnum (Figure 2A). At the cervicothoracic level, MRI showed a cervicothoracic syringomyelia cavity initially interpreted by the radiologist in a satellite center as a medullary cystic infiltrating tumor because of the associated medullary oedema (Figure 2A).

A diagnosis of Chiari type I malformation complicated by hydrocephalus and cervicothoracic syringomyelia was made in view of the observed radiological lesions. A PCMRI analysis was not performed preoperatively because of the patient's clinical condition and the need to perform an emergency procedure. Therefore, the hydrocephalus was initially treated by endoscopic ventriculocisternostomy.

The postoperative course was marked by the regression of the vigilance disorders, the disappearance of the headaches, and the disappearance of the oculomotor disorders. There remained only a few sensitivity disorders such as paraesthesia of the limbs, which disappeared at 2 months postoperatively. Close monitoring was proposed, with a follow-up MRI at 3 weeks combining phase contrast sequences.

MRI showed a complete reduction in cerebellar tonsil ptosis and a decrease in ventricular volume (Figures 1B, 1C, 2B, and 2C). Phase contrast MRI showed, at 3 weeks postoperatively, a stenosis of the midbrain aqueduct and complete disappearance of the syringomyelia. The pulsatility of the CSF at the endoscopic third ventriculostomy was significant (345 microliters/cardiac cycle), as well as a hyperpulsatility of the subarachnoid CSF measured intracranially (at the level of the prepontic cisterns) and at the cervical level (at the C2C3 disc). This pulsatility was 953 microliters per cardiac cycle at the cervical level (follow-up value: 340 +- 210 microliters per cardiac cycle) [10]. This hyperpulsatility of the CSF responded to increased intracranial vascular volume variation (1.5 mL/ cardiac cycle) (control value: 0.76+-0.30 mL/ cardiac cycle) [10].

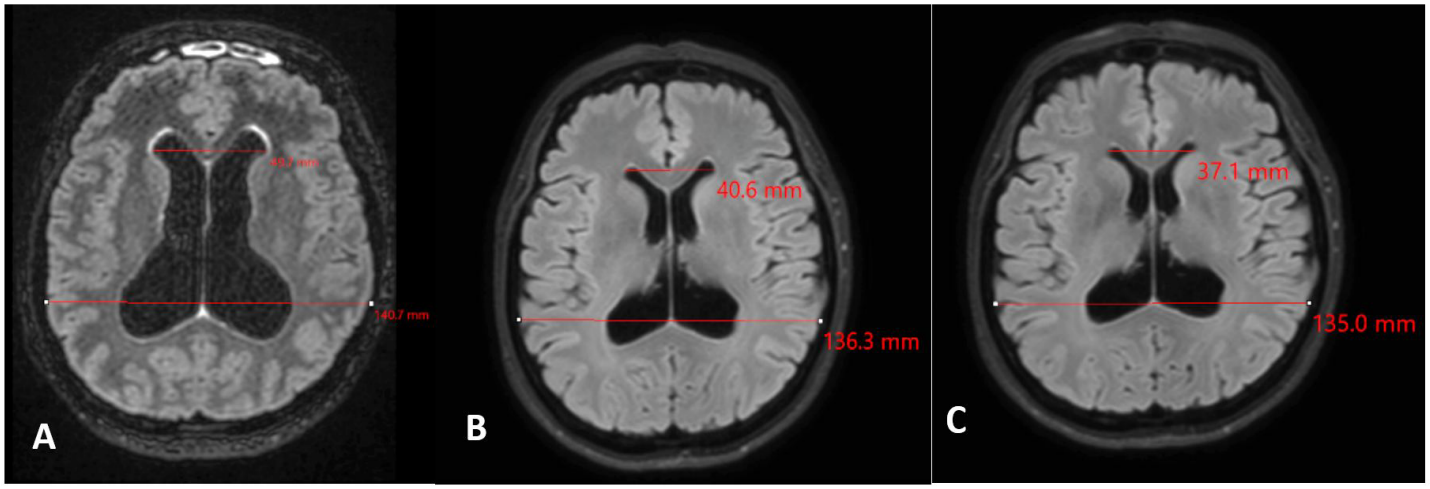

Figure 1: Axial MRI scans showing the resolution of the hydrocephaly. (A) Axial MRI scan before surgery showing hydrocephalus with Evans index to 0.35. (B) Axial FLAIR MRI at 3-month post-surgery with Evans index to 0.29. (C) Axial FLAIR MRI at 19-month post-surgery with Evans index to 0.27.

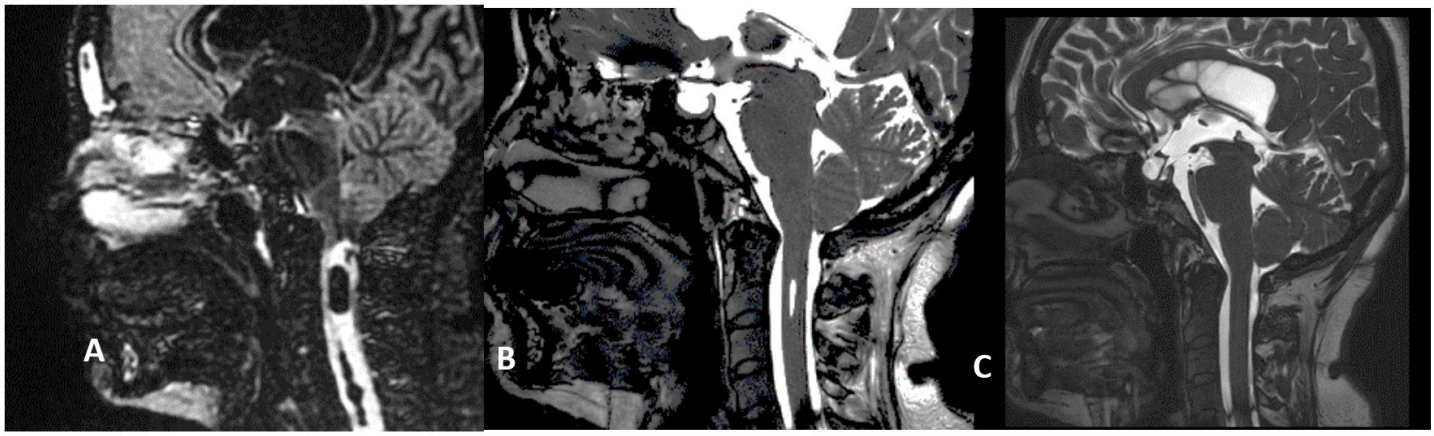

Figure 2: Sagittal MRI view showing the resolution of syrinx. (A) MRI T2 FLAIR pre operative showing the syrinx at the cervical spinal cord level. (B) T2 MRI at 3 weeks after surgery with regression of syringomyelia. (C) T2 MRI at 12 months after surgery showing disappearance of the syringomyelia.

Discussion

Our clinical case shows that the diagnosis of Chiari malformation can be secondary to another pathology, such as hydrocephalus. Our patient presented with pre-existing hydrocephalus, probably previously compensated, which generated an increasingly high pressure gradient between the supratentorial and infra-tentorial spaces first. Secondly, the supratentorial hydrocephalus caused an increase in infratentorial pressure that resulted in chronic tonsillar involvement.

The pathophysiology of hydrocephalus associated with MCI is debated [5-7, 11]. For some authors, the Chiari anomaly is responsible for a distal obstruction of the outflow tracts at the level of V4 (foramen of Magendie and Lushka). For other authors, it could be an obstruction of the outflow tracts of V4, which would cause ptosis of the cerebellar tonsils. In the series of Decq [6], it has been shown that ventriculocisternostomy was very effective in hydrocephalus associated with MCI. In this series, it was essentially MCI associated with an obstruction of Magendie's foramen. A partial regression of the degree of tonsil ptosis was observed postoperatively. In a broad review of the literature, Massimi et al. [12] proposed multiple etiologies for hydrocephalus associated with a Chiari anomaly. However,

he found a predominance of distal obstruction mechanisms at the level of V4 by the MCI justifying the indications of endoscopic ventriculocisternostomy. Only 11% of patients treated by ETV underwent posterior fossa decompression because of persistent symptoms.

Our clinical case shows the usefulness of PCMRI in the understanding of the physiopathological mechanisms of MCI. In our clinical case, in view of the life-threatening hydrocephalus, it was not possible to perform a PCMRI to study the hemodynamic changes at the level of the cranio-spinal hinge. The postoperative PCMRI was able to determine the etiology of the hydrocephalus by revealing an absence of flow at the aqueductal level, indicating a stenosis. Moreover, on the later follow-up examinations, it was found that the ptosis of the tonsils had completely regressed. This supports the existence of a MCI, which would be secondary to hydrocephalus rather than the opposite. The etiology would rather be that of tonsillar involvement generated by a pressure gradient than a MCI. Performing a PCMRI at the diagnostic phase allows a comprehensive analysis of craniospinal hemodynamics and hydrodynamics [13]. In cases where MRI can be performed before the procedure, it enables confirmation of the therapeutic approach.

The therapeutic approach to hydrocephalus associated with MCI is also debated. A large body of literature suggests that ETV should be performed [5, 6, 12]. Some authors have suggested a posterior fossa decompression [11]. ETV has a low complication rate [12] and high clinical effectiveness. Decq et al. [6], showed a regression of the degree of preoperative tonsil ptosis from 13.75 mm to 7.76 mm post-surgery. This reflects the effectiveness of sustentorial hydraulic treatment of the pathophysiological context as a whole.

Our patient had significant intracranial vascular volume variation in view of the data in the control populations (approximately twice as high) [10]. The vascular expansion is compensated by flushing of CSF into the perimedullary subarachnoid spaces [8, 9]. In patients with a Chiari anomaly associated with hydrocephalus, the hydrocephalus may aggravate the degree of ptosis of the cerebellar tonsils and thus lead to a decrease in the CSF outflow area at the foramen magnum. A decrease in the flow area leads to an increased resistance to flow, thus altering the pulsatility of the CSF at this level. The alteration of this mobile compliance would lead to an exponential worsening of the symptomatology due to the loss of one of the main mechanisms of intracranial pressure self-regulation. Performing an ETV would make it possible to reduce or even eliminate the pressure gradient. This gradient would be at the origin of the increased ptosis of the tonsils and thus of the restriction of the surface of flow of the CSF at the level of the foramen magnum. In this way, the Chiari anomaly is not treated, but the mechanisms of compensation and regulation of the intracranial pressure would be restored. In our case, this resulted in a clear symptomatic improvement and a nearly complete regression of the syringomyelia cavity.

In our case, the ptosis of the cerebellar tonsils is not a MCI but a tonsillar commitment by the installation of a pressure gradient between the intracranial and perimedullary subarachnoid spaces. This mechanism is quite similar for intracranial expansive processes, lumboperitoneal shunts, lumbar CSF leaks [14, 15].

Conclusion

The pathophysiology of Chiari anomalies associated with hydrocephalus could be clarified by performing a PCMRI. Carrying out this examination could thus improve our diagnostic ability and thus adapt the therapy to the particular context of the patient. ETV seems the therapeutic option to reduce the hydrodynamic consequences of hydrocephalus and its impact on MCI.

References

1. Aboulezz AO, Sartor K, Geyer CA, Gado MH (1985) Position of Cerebellar Tonsils in the Normal Population and in Patients with Chiari Malformation: A Quantitative Approach with MR Imaging, Journal of Computer Assisted Tomography, 9: 1033-1036.
2. Aitken LA, Lindan CE, Sidney S, Gupta N, Barkovich AJ, et al. (2009) Chiari Type I Malformation in a Pediatric Population, Pediatric Neurology; 40: 449-454.
3. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, et al. (1999) Chiari I Malformation Redefined: Clinical and Radiographic Findings for 364 Symptomatic Patients. Neurosurgery, 44: 1005-1017.
4. EA Bering Jr (1962) Circulation of the Cerebrospinal Fluid: Demonstration of the Choroid Plexuses as the Generator of the Force for Flow of Fluid and Ventricular Enlargement. Journal of Neurosurgery, 19: 405-413.
5. Massimi L, Pravatà E, Tamburrini G, Gaudino S, Pettorini B, et al., (2011) Endoscopic Third Ventriculostomy for the Management of Chiari I and Related Hydrocephalus: Outcome and Pathogenetic Implications, Neurosurgery, 68: 950-956.
6. Sainte-Rose C, Cinalli G, E. Roux F, Maixner W, Chumas PD, et al. (2001) Third Ventriculostomy (2001) Journal of Neurosurgery, 95.
7. Mohanty A, Suman R, SR Shankar, S Satish, SS Praharaj (2005) Endoscopic third ventriculostomy in the management of Chiari I malformation and syringomyelia associated with hydrocephalus, Clinical Neurology and Neurosurgery, 108: 87-92.
8. O Balédent, MC Henry-Feugeas, I Idy-Peretti (2001) Cerebrospinal Fluid Dynamics and Relation with Blood Flow: A Magnetic Resonance Study with Semiautomated Cerebrospinal Fluid Segmentation, Investigative Radiology, 36: 368-377.
9. D Greitz (2004) Radiological assessment of hydrocephalus: new theories and implications for therapy, Neurosurg Rev, 27: 145-165.
10. A Lokossou, S Metanbou, C Gondry-Jouet, O Balédent (2020) Extracranial versus intracranial hydro-hemodynamics during aging: a PC-MRI pilot cross-sectional study, Fluids Barriers CNS, 17.
11. J Koueik, RL DeSanti, BJ Iskandar (2022) Posterior fossa decompression for children with Chiari I malformation and hydrocephalus, Childs Nerv Syst, 38: 153-161.

12. L Massimi, G Pennisi, P Frassanito, G Tamburrini, C Di Rocco, M Caldarelli (2019) Chiari type I and hydrocephalus, Childs Nerv Syst, 35: 1701-1709.

13. C Capel, P Padovani, PH Launois, S Metanbou, O Balédent, J Peltier (2022) Insights on the Hydrodynamics of Chiari Malformation, JCM, 11: 5343.

14. TH Milhorat, M Nishikawa, RW Kula, YD Dlugacz (2010) Mechanisms of cerebellar tonsil herniation in patients with Chiari malformations as guide to clinical management, Acta Neurochir, 152: 1117-1127.

15. T Morioka, T Shono, S Nishio, K Yoshida, K Hasuo, M Fukui (1995) Acquired Chiari I malformation and syringomyelia associated with bilateral chronic subdural hematoma: Case report, Journal of Neurosurgery. 83: 556-558.