www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, NYU Winthrop Hospital, Mineola, NY, USA.

SNI: General Neurosurgery

Editor
Eric Nussbaum, MD
National Brain Aneurysm and Tumor Center, Twin Cities, MN, USA

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

SNI
Open Access

Case Report

Acquired Chiari I malformation due to lumboperitoneal shunt: A case report and review of literature

Aslam Hentati, Mohamed Badri, Kamel Bahri, Ihsen Zammel

Department of Neurosurgery, Burns and Trauma Center, Ben Arous, Tunis, Tunisia.

E-mail: *Aslam Hentati - aslamhentati@gmail.com; Mohamed Badri - badri.med@gmail.com; Kamel Bahri - kamelbahri@yahoo.com; Ihsen Zammel - ihsenzammel@gmail.com

***Corresponding author:**
Aslam Hentati,
Department of Neurosurgery,
Burns and Trauma Center,
Ben Arous, Tunis, Tunisia.

aslamhentati@gmail.com

Received : 17 December 18
Accepted : 04 April 19
Published : 10 May 19

DOI
10.25259/SNI-234-2019

Quick Response Code:

ABSTRACT

Background: The Type I malformations are supposed to be the result of mesodermal defects that create a congenitally small posterior fossa. However, Chiari malformation could be also "iatrogenic" and then called "acquired" Chiari I malformation. In this study, the authors report the clinical feature of a patient who developed a Chiari I malformation after lumboperitoneal shunt.

Case Description: A 35-year-old woman has been suffering from idiopathic intracranial hypertension and rhinorrhea due to an anterior skull base defect. A valveless lumboperitoneal shunt followed by surgical closure of the defect was performed. Six months later, she suffered from major continuous occipital headaches. The neurological examination found a mild cerebellar gait ataxia and cerebellar dysarthria. The cerebral magnetic resonance imaging (MRI) showed a ptosis of the cerebellar tonsils and a disappearance of the cisterna magna; there was no syringomyelia. This herniation was not present before shunt was performed. A replacement of the lumboperitoneal shunt with a pressure-regulated valve chamber was performed. After a 1-year follow-up, the patient reports a marked decrease of the headache as well as the ataxia, and the last cerebral MRI showed resolution of the Chiari I malformation.

Conclusions: Symptomatic acquired Chiari malformation with or without syringomyelia as a delayed complication after lumbar shunting is a rare complication, particularly reported in the pediatric population, but could also occur to adult patients. Treating these patients by correcting the shunt's valve could be enough, but should be monitored, as it may fail to resolve the Chiari malformation even years after treatment.

Keywords: Chiari malformation, Complication, Lumbar shunt

INTRODUCTION

The Chiari I malformation was first described by Chiari, in 1891.[2] It is characterized by the presence of cerebellar tonsils in the upper cervical canal while the fourth ventricle remains above the foramen magnum.[2] The Type I malformations are supposed to be the result of mesodermal defects that create a congenitally small posterior fossa.[12] However, Chiari malformation could be also "iatrogenic" and then called "acquired" Chiari I malformation.[12,21] In fact, the term of acquired Chiari malformation (ACM) was used by Payner *et al.* in the presence of the herniation of cerebellar tonsils subsequent to the documentation of a normal hindbrain in prior brain imaging.[14] It was mainly described after cerebrospinal fluid (CSF) shunting, first after lumboperitoneal shunting, and lately recognized after supratentorial shunting.[1,3,5,14]

In this study, we report the clinical features of a patient who developed a Chiari I malformation after lumboperitoneal shunt.

CASE REPORT

A 35-year-old woman has been suffering from idiopathic intracranial hypertension for 8 years. For 3 years, the patient has had rhinorrhea due to an anterior skull base defect. A valveless lumboperitoneal shunt followed by surgical closure of the defect was performed. Six months later, she suffered from major continuous occipital headaches with nauseas. The neurological examination found a mild cerebellar gait ataxia with limb incoordination and cerebellar dysarthria. Cranial nerve examination was normal. The cerebral magnetic resonance imaging (MRI) showed a ptosis of the cerebellar tonsils and a disappearance of the cisterna magna [Figure 1]. This herniation was not present before shunt was performed [Figure 2]. A replacement of the lumboperitoneal shunt with a pressure-regulated valve chamber was implemented. After a 1-year follow-up, the patient reports a marked decrease of the headache as well as the ataxia. The last cerebral MRI showed resolution of the Chiari I malformation [Figure 3].

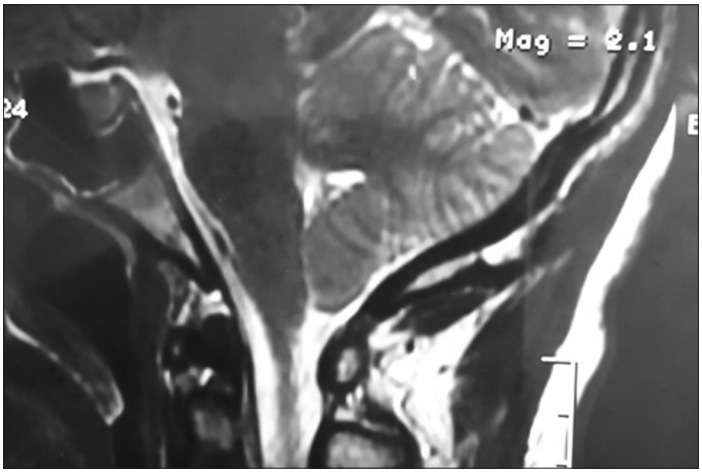

Figure 1: After performing a valveless lumboperitoneal shunt, the cerebral MRI in sagittal section showed an acquired Chiari malformation.

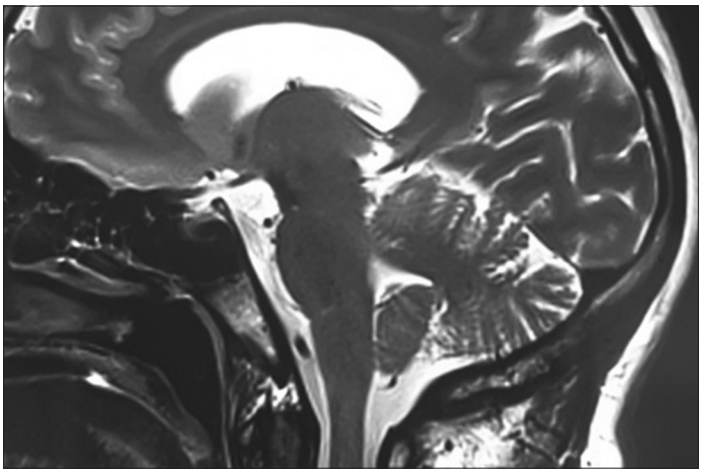

Figure 2: Preoperative cerebral MRI in sagittal section. There was no Chiari malformation.

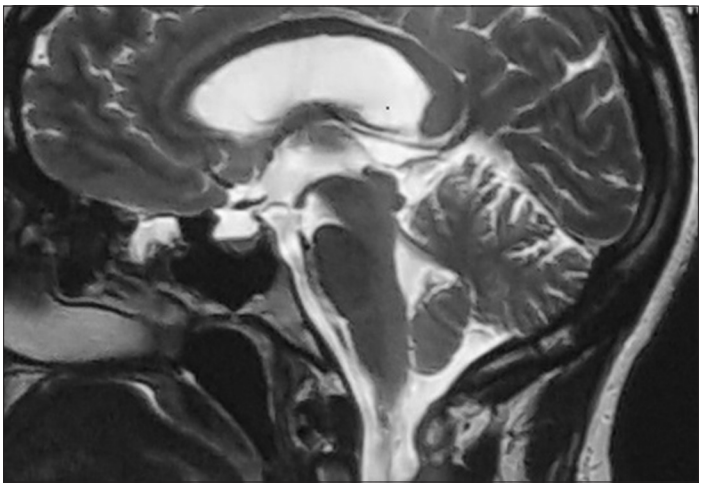

Figure 3: After putting a pressure-regulated lumboperitoneal shunt valve, the cerebral magnetic resonance imaging in sagittal section showed a reascending of the cerebellar amygdala.

DISCUSSION

Chiari malformation type I (CM-I) has traditionally been defined as a dislocation of the cerebellar tonsils 5 mm or more below the foramen magnum on sagittal MRI.[11] CM-I is often associated with basilar invagination, as described by Liao *et al*., as they represent the most common craniovertebral junction malformations (CVJ).[10] Furthermore, they can be associated with other miscellaneous CVJ malformations such as platybasia or more rarely a defect of closure of the posterior arch of C1, an occipitalization of the atlas, a C1–C2 dislocation, and C1–C2 fusion.[4,10]

Unlike the "classic" CM-I, ACM is defined by the herniation of cerebellar tonsils subsequent normal hindbrain in prior brain imaging, and it is not related to CVJ malformations.[14] ACM was first described by Hoffman and Tucker, in 1976, as a late complication of lumboperitoneal shunt.[7] To the best of our knowledge, 36 patients of ACM due to lumboperitoneal (LP) shunts have been reported since 1976.[3,7,8,13-15,17,18,21] Among them, 27 cases are summarized in the table below. These patients were selected as they present enough information about their imaging (MRI and/or computed tomography scan), clinical data, and postoperative outcome.

The incidence of ACM due to LP shunt varies in different series. Chumas *et al*. reviewed a series of 143 patients to determine the incidence of ACM and found that the incidence of hindbrain herniation may be as high as 70% in asymptomatic patients with LP shunts.[3] Five of these patients became symptomatic and required Chiari decompression.[3] During their follow-up period, which is about 5.7 years, there was one shunt-related death due to unsuspected tonsillar herniation.[3] However, in two other series of 25 patients by Rekate and Wallace and of 74 patients by Wang, no patient developed an ACM.[16] In another series of 70 patients treated with LP shunting for pseudotumor cerebri, Johnston *et al*. reported an incidence of an ACM in 11 patients.[8] Eight of them

Table 1: Reported cases of acquired Chiari malformation due to lumboperitoneal shunt.

Author	Patient number	Age (Y)	Sex	Diagnosis	Method	Shunt type	Time to sympt. (y)	Syringom	Symptoms	Treatment	Clinical outcome	Imaging outcome
Sullivan *et al.*, 1988[18]	1	36	Female	Pseud.	MR imaging	LP	4	Yes	Headaches, weakness of the left arm	LP to VA	Resolved	Resolved
Chumas *et al.*, 1993[3]	2	0.5	n/a	Crouz's des.	Not specified	LP	1.7	No	Neck stiffness, fever, cardiorespiratory arrest	LP revision then ACM decompression	Died	Died
	3	0.7	n/a	Hydroc.	Not specified	LP	3.4	Yes	Quadriparesis with spasticity	ACM decompression	Resolved	Persistent
	4	0.5	n/a	Hydroc.	Not specified	LP	5.4	Yes	Gait ataxia, hemiparesis	LP To VP then ACM decompression	Resolved	Persistent
	5	1.5	n/a	Hydroc.	Not specified	LP	0.6	No	Neck pain, ataxia, arm weakness	ACM decompression	Resolved	Persistent
	6	0.5	n/a	Hydroc.	Not specified	LP	8.7	No	Suboccipital headache, neck stiffness	ACM decompression	resolved	Persistent
	7	0.75	n/a	Hydroc.	Not specified	LP	16.6	No	Suboccipital headache, neck stiffness	ACM decompression	Resolved	Persistent
Payner *et al.*, 1994[12]	8	1.25	Male	Hydroc.	MR imaging	LP	1.1	No	Head tilt, truncal ataxia	LP to VP	Resolved	
	9	1	Female	Hydroc.	MR imaging	LP	0.75	no	Apnea, stridor	LP to VP	Resolved	
	10	7	Female	Pseud.	MR imaging	LP	2	No	Headache	Posterior fossa decompression	Resolved	
	11	5	Male	Pseud.	MR imaging	LP	5	No	Headache, leg paresthesis	Posterior fossa decompression	Resolved	
Johnston *et al.*, 1998[8]	12	Child	Female	Crouz's des.	Not specified	LP	19	Yes	Limb weakness	Syrinx to SAS shunt	Some improvement	
	13	Child	Male	Crouzon's des.	MR imaging	LP	4	Yes	Leg weakness	LP to VP shunt	Resolved	Syrinx resolved
	14	Child	Female	Apert's	Not specified	LP	3	Yes	Bilateral leg weakness	Syrinx to SAS shunt then ACM decompression	Gradually worse then improved	Improved
	15	Child	Male	Hydrocephalus	MR imaging	LP	4.5	No	Severe interscapular pain	LP to VP	Resolved	
	16	Child	Male	Hydrocephalus	MR imaging	LP	1	Yes	Drowsiness, bilateral motor signs	ACM decompression HV valve to the LP shunt	Resolved	

(Contd...)

Table 1: *Continued*

Author	Patient number	Age (Y)	Sex	Diagnosis	Method	Shunt type	Time to sympt. (y)	Syringom	Symptoms	Treatment	Clinical outcome	Imaging outcome
	17	12	Female	Hydrocephalus	MR imaging	LP	16	No	Neck pain, cranial nerve palsy, ataxia	ACM decompression then LP to VA	Worse then resolved	
	18	Child	Male	Hydrocephalus	MR imaging	LP	9	Yes	Headache neck pain	LP to VP then ACM decompression	Persisted then resolved	
	19	Child	Female	Intracranial hyp.	MR imaging	LP	3	Yes	Disco sensory loss	Syrinx to SAS shunt	Improved	
		Child					7	Yes	Dizziness, neck pain	ACM decompression	Resolved	
	20	Child	Female	Intracranial hyp.	MR imaging	LP	2.5	No	Headache, nausea, ataxia	LP to CA	Resolved	
	21	Child	Fenale	Intracranial hyp.	MR imaging	LP	3	No	Headache, ataxia	LP to CA then ACM decompression	Recurred then resolved	
	22	Child	Female	Occipital PM	MR imaging	LP	5	Yes	Interscapular pain	LP to VP	Resolved	
	23	Child	Female	Lumbar PM	MR imaging	Cystopertneal	9	Yes	Discosensory loss	Syringopleural shunt	Improved	
Padmanabhan *et al.*, 2005[11]	24	31	Female	Intracranial hyp.	MR imaging	LP	0.5	Yes	Hemisensory loss, left-hand motor deficit	LP to VP	Resolved	Persistent
Riffaud *et al.*, 2008[17]	25	1.5	Male	Gorlin's syn.	MR imaging	LP (valveless)	11.5	Yes	Headaches	Valveless LP shunt to Valved LP shunt	Resolved	Resolved
Peleggi and Lovely, 2012[13]	26	15	Female	Intracranial hyp.	MR imaging	LP	10	Yes	Headaches, vomiting, sensory loss	LP to VP then ACM decompression	Persisted then resolved	Persistent then resolved
Current case	27	35	Female	Intracranial hyp.	MR imaging	LP	1	No	Headaches	Pressure-regulated valve to the LP shunt	Resolved	Resolved

Y: Years, n/a: Not available, Pseud.: Pseudotumor cerebri, Crouz's dis.: Crouzon's disease, Hydroc.: Hydrocephalus, LP: Lumboperitoneal, VP: Ventriculoperitoneal, VA: Ventriculoatrial, Intracranial hyp.: Intracranial hypertension, MR imaging: Magnetic resonance imaging, Time to sympt.: Time delay between shunt implantation and clinical signs, Syring.: Syringomyelia

were asymptomatic and three required treatment.[8] Despite the variability of these results, ACM due to LP shunting should be considered, even if it is mostly asymptomatic. However, it should require a long follow-up due to the possible late onset of the symptomatology.[3,8]

All reported patients with ACM were children, except for the case described in this paper and the one reported by Padmanabhan *et al*.[13] Most of the symptoms were a manifestation of chronic tonsillar herniation (21 patients; 80%) (i.e., occipital and posterior cervical pain, long-track deficits, lower cranial nerve palsies, brain stem dysfunction, and cerebellar syndrome), whereas symptoms related to syringomyelia were less frequent (i.e., sensory loss or weakness in upper limbs).

ACM with or without syringomyelia had been noted after lumbar shunting regardless of the underlying etiology[8] [Table 1]. It was mostly described in cases of pseudotumor cerebri and communicating hydrocephalus.[3,7,8,13-15,17,18,21]

While the cause of the ACM is debatable, most authors consider that it is a result of CSF pressure differentials across the cranial cervical junction, created by the drainage.[8,12,15] According to Johnston *et al*., the pathogenesis of syrinx formation may be explained by the hydrodynamic theory, as the lumbar shunt can be responsible of the disorders of CSF circulation.[8] Johnston *et al*. also reported one patient (number 14 in the table) having syrinx formation before ACM suggesting that "Chiari malformation may follow rather precede syrinx formation" in some cases.[8]

The "classic" CM-I is being increasingly diagnosed, but its optimal surgical treatment remains controversial.[9,19,20] Transoral odontoidectomy followed by occipitocervical fixation is a used approach to relieve ventral compressions.[6,20] Congenital CM-I is most treated by surgical posterior fossa decompression.[7,19] The traditional technique was to perform a suboccipital craniotomy and creating an artificially enlarged cisterna magna, suturing dural edges to the divided suboccipital muscles with wide arachnoid dissection.[22] A modification of this technique includes dural plasty, or with dura splitting or bone-only decompression.[9,19] Suboccipital craniotomy and opening the dura without opening the arachnoid, as described by Lavorato *et al*., is trending to be a good surgical strategy that can lead to an improvement of syringomyelic cavities.[9]

As for ACM due to LP shunt, the number of reported cases (27) is too limited to make clear and undoubtful therapeutic recommendations, but three possible strategies have been described:

1. To put a valve to a valveless shunt.
2. To resite the shunt.
3. To decompress the craniocervical junction.

Putting a valve to an LP shunt or resiting it could be an efficient solution to isolated ACMs. However, these techniques could not be enough when ACM is associated with syringomyelia, as symptoms did not resolve or could even get worse after a short or long follow-up. Craniocervical junction's decompression could be required days or even years after shunt revision. It is also reported that isolated ACM could be treated by posterior fossa decompression alone, with good clinical and radiological outcome. Thus, a good and long follow-up after surgery is required, as a clinical relapse could lead to recite the shunt. Sullivan *et al*. and Riffaud *et al*. (patients number 1 and 25) reported a good recovery of patients having an ACM associated with syringomyelia after putting a valve to an LP shunt or resisting it.[17,18] However, the follow-up was not long enough (6 and 18 months), as a relapse could happen years after initial treatment (patients number 17 and 19).[8,17,18] Putting a pressure-regulated valve for the case reported in this paper is believed to be efficient. It was an isolated ACM, and symptoms resolved with a reascension of cerebellar tonsils. However, longer follow-up could also be required like for the other reportedly treated ACMs.

CONCLUSIONS

Symptomatic ACM, a rare complication of lumbar shunt, is particularly described in the pediatric population but could also occur to adult patients. The review of these reported cases suggests that the association with syringomyelia could be decisive about the need of decompression of the craniocervical junction.

Patients treated by only correcting the lumbar shunt should be monitored, as a relapse of the complication could happen years after initial treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Caldarelli M, Novegno F, Di Rocco C. A late complication of CSF shunting: Acquired Chiari i malformation. Childs Nerv Syst 2009; 25:443-52.
2. Chiari H. Ueber veränderungen des kleinhirns infolge von hydrocephalie des grosshirns. Dtsch Med Wochenschr 1891;42: 1172-5.
3. Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ,

Humphreys RP, *et al*. Tonsillar herniation: The rule rather than the exception after lumboperitoneal shunting in the pediatric population. J Neurosurg 1993;78:568-73.
4. Ciappetta P, Signorelli F, Visocchi M. The role of arachnoid veils in chiari malformation associated with syringomyelia. Acta Neurochir Suppl 2019;125:97-9.
5. Di Rocco C, Velardi F. Acquired chiari Type l malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 2003; 19:800-7.
6. Dickman CA, Locantro J, Fessler RG. The influence of transoral odontoid resection on stability of the craniovertebral junction. J Neurosurg 1992;77:525-30.
7. Hoffman HJ, Tucker WS. Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. Childs Brain 1976;2:167-76.
8. Johnston I, Jacobson E, Besser M. The acquired chiari malformation and syringomyelia following spinal CSF drainage: A study of incidence and management. Acta Neurochir (Wien) 1998;140:417-27.
9. Lavorato L, Spallone A, Visocchi M. Surgical treatment of chiari malformation in adults: Comparison of surgical techniques described in the literature and our experience. Acta Neurochir Suppl 2019;125:139-43.
10. Liao C, Visocchi M, Zhang W, Li S, Yang M, Zhong W, *et al*. The relationship between basilar invagination and chiari malformation Type I: A narrative review. Acta Neurochir Suppl 2019;125:111-8.
11. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, *et al*. Chiari i malformation redefined: Clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 1999;44:1005-17.
12. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y. Pathogenesis of chiari malformation: A morphometric study of the posterior cranial fossa. J Neurosurg 1997;86:40-7.
13. Padmanabhan R, Crompton D, Burn D, Birchall D. Acquired chiari 1 malformation and syringomyelia following lumboperitoneal shunting for pseudotumour cerebri. J Neurol Neurosurg Psychiatry 2005;76:298.
14. Payner TD, Prenger E, Berger TS, Crone KR. On the pathogenesis of syringomyelia: A review. J R Soc Med 1994;73:798-806.
15. Peleggi AF, Lovely TJ. Treatment of delayed chiari malformation and syringomyelia after lumboperitoneal shunt placement: Case report and treatment recommendations. Surg Neurol Int 2012;3:101.
16. Rekate HL, Wallace D. Lumboperitoneal shunts in children. Pediatr Neurosurg 2003;38:41-6.
17. Riffaud L, Moughty C, Henaux PL, Haegelen C, Morandi X. Acquired chiari i malformation and syringomyelia after valveless lumboperitoneal shunt in infancy. Pediatr Neurosurg 2008;44:229-33.
18. Sullivan LP, Stears JC, Ringel SP. Resolution of syringomyelia and chiari i malformation by ventriculoatrial shunting in a patient with pseudotumor cerebri and a lumboperitoneal shunt. Neurosurgery 1988;22:744-7.
19. Villa A, Imperato A, Maugeri R, Visocchi M, Iacopino DG, Francaviglia N. Surgical treatment in symptomatic chiari malformation Type I: A series of 25 adult patients treated with cerebellar tonsil shrinkage. Acta Neurochir Suppl 2019;125:125-31.
20. Visocchi M, Trevisi G, Iacopino DG, Tamburrini G, Caldarelli M, Barbagallo GM. Odontoid process and clival regeneration with chiari malformation worsening after transoral decompression: An unexpected and previously unreported cause of "accordion phenomenon." Eur Spine J 2015;24:564-8.
21. Welch K, Shillito J, Strand R, Fischer EG, Winston KR. Chiari i "malformations" an acquired disorder? J Neurosurg 1981;55:604-9.
22. Williams B. Surgery for hindbrain related syringomyelia. Adv Tech Stand Neurosurg 1993;20:107-64.

How to cite this article: Hentati A, Badri M, Bahri K, Zammel I. Acquired Chiari I malformation due to lumboperitoneal shunt: A case report and review of literature. Surg Neurol Int 2019;10:78.