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Original article

Chiari malformations in adults: A single center surgical experience with special emphasis on the kinetics of clinical improvement



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ARTICLE INFO

Article history:

Received 21 October 2017

Received in revised form 26 August 2018

Accepted 6 October 2018

Available online 8 February 2019

Keywords:

Chiari malformation

Occipito-cervical decompression -

Syringomyelia

ABSTRACT

Background. – The Chiari malformation type I (CM-I) is the most commonly found type in adults. The efforts to further improve the treatment offered for these malformations are hampered by the existence of controversial methods and the absence of a uniform scoring system to evaluate clinical outcomes.

Objectives. – The goal of our study is to analyze the clinical and radiological data concerning patients operated for CM and to expose surgical techniques.

Patients and treatment. – This is a retrospective study concerning patients of more than 16 years of age, operated on (from 2000 to 2016) in our institution. These patients underwent bony decompression of the cervico-occipital junction, with a duraplasty enlargement. Clinical and radiological follow-up was assessed sequentially.

Results. – The mean age of patients included in this study was 39. Headaches ($n = 19$) and sensory disturbances ($n = 17$) were the most common presenting complaints. Furthermore, syringomyelia symptoms were present in 34.5% of the cases ($n = 10$). Twenty-three patients displayed a Chiari malformation of type I (79.3%), and six patients were classified as Chiari malformation type 1.5 (20.7%). A syringomyelia was present in 58.6% of the cases ($n = 17$). The postoperative complications that were encountered were: one case of pseudo-meningocele, two cases of cerebrospinal fluid leakage, two cases of meningitis, and one case of delayed wound healing. The mean follow-up period was 18 months, which showed beneficial outcomes in 82.8% of the cases (20.7% cured, and 62.1% improved) and an unfavorable outcome in 17.2% of the cases (13.8% stable patients and 3.4% worsened outcomes). Syringomyelia symptoms were improved in 60% of the patients. Among the patients who presented without a syrinx, 82.3% had good outcomes; and those who presented with a syrinx, 83.4% had good outcomes. Symptoms improved for 69% of patients within 3 months.

Conclusion. – An optimal craniocervical osteo-dural decompression plus duraplasty offered early and sustainable good clinical results in symptomatic CM-I and CM-I.5 patients.

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1. Introduction

The Chiari malformation is a rare congenital nervous system malformation that consists of a downward herniation of the caudal portion of the cerebellum and sometimes below the foramen magnum [1]. It must be differentiated from cerebellar amygdala herniation [2]. The most common type of CM is type I, it can be latent for a long time, and is primarily discovered in

adulthood. It can present with several clinical manifestations and some complications resulting from the repercussions of the malformation on the brainstem, the inferior part of the cerebellum or the superior part of the spinal cord [2]. An additional threat is the development of syringomyelia resulting in an anomaly in the normal circulation of the cerebrospinal fluid (CSF) at the level of the foramen magnum.

The management of a symptomatic or worsening malformation is surgical. This approach aims to facilitate the CSF circulation. Different techniques have been developed, spanning from simple duplication of the dura to a more complex opening and tonsillar resection. In order to investigate the craniocervical bony

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decompression and the duraplasty surgery for CM type I patients, the results and outcome of a series of 29 patients operated with this technique were analyzed.

2. Patients and methods

Our study is a retrospective analysis of the clinical, radiological, and therapeutic data concerning patients of more than 16 years of age, admitted to neurosurgery for CM. The diagnosis was approved for herniation of the cerebellar amygdala of more than or equal to 5 mm in relation to the line of Mc Rae [3]. Excluded from this study were all the abnormal cerebellar amygdala positions in relation to the process of engagement. All 29 patients underwent an occipitocervical decompression through a posterior approach.

We classified our patients after surgery into four outcome categories, and for each patient the classification of a particular outcome category was based on the evolution of the principal signs and symptoms which included: healing (complete remission or marked improvement), good evolution (slight improvement), stable evolution (stable signs and symptoms), unfavorable evolution or aggravation (slight or marked deterioration).

2.1. Description of the surgical technique

We performed an occipital craniectomy of 4 × 4 cm, associated with ablation of the posterior arch of C1, sometimes extending to C2 and then suspending the dura matter after opening it in a “Y”. Then an enlargement plasty is sutured with synthetic dura (neuropatch) suspended from the muscular layer.

2.2. Postoperative follow-up protocol

On average, patients are discharged 8 days after surgery, the first MRI was performed at 3 months and an additional MRI with flow sequence was done at 12 months. Patients were then clinically checked annually (Fig. 1).

3. Results

The average age of our patients was 39 years old, ranging from 19 to 66 years of age. The most affected age group was between 19 to 29 years old (31.1%). We noted a slight female predominance with 58.6% against 41.4% in the males (Sex ratio = 0.7; 12 males; 17 females). We reported a case of CM decompensated by cervical trauma after a water sport. The remainder of the patients had no previous medical or surgical history in relation to our study. All patients were symptomatic. Headache and particularly those linked to paroxysmic intracranial hypertension, neck pain and sensory symptoms are cited as the most common presenting complains. Clinical signs of syringomyelia were present in 10 cases (34.5%) (Table 1).

In all cases, MRI has confirmed the diagnosis of CM. Twenty-one patients (79.3%) had type I CM and 6 patients (20.7%) were categorized CM of type I.5 which corresponds to a hernia of the cerebellar tonsils and the medulla oblongata. Syringomyelia was present on MRI in 58.6% of cases ($n = 17$) and the most frequent location being cervical (41.2%). Syringomyelia was pan-medullary in 35.3% of cases, cervico-dorsal in 17.6% of cases and pure dorsal in 5.9% of cases.

As for the postoperative complications, we encountered one case of pseudo-meningocele which did not necessitate any treatment, two cases of CSF leakage, in which one of them needed to be reoperated. The second case of CSF leakage necessitated a lumbar shunt to be placed which had resolved the problem. One case of delayed wound healing was noted and two cases of meningitis were

Table 1

Presenting signs and symptoms of 29 adult patients with CM.

Symptoms and signs	Number of patients (%)
Headaches	19 (65.5%)
Neck pain ^a	14 (48.3%)
Sensory symptoms ^a	17 (58.6%)
Motor symptoms ^a	8 (27.6%)
Dizziness	3 (10.3%)
Discomfort	3 (10.3%)
Dysphagia	2 (6.9%)
Tinnitus	2 (6.9%)
Vomiting	1 (3.4%)
Sphincter dysfunction	1 (3.4%)
Pyramidal syndrome	6 (20.7%)
Cerebellar syndrome	3 (10.3%)
Posterior cord syndrome	1 (3.5%)
Syringomyelic syndrome ^a	10 (34.5%)
Cranial nerve impairment	2 (6.9%)
Nystagmus	1 (3.4%)
Scoliosis	1 (3.4%)

^a Symptoms may also be due to the presence of the syringomyelic cavity created by the CM.

treated by antibiotic therapy. The neurological evolution was evaluated in the early postoperative phase and on a regular basis. The postoperative follow-up period ranged from 3 months to 8 years with a mean follow-up of 20 months and an average follow-up of 26 months.

The results of our patients during the last consultation are summarized in (Fig. 2). Sixty nine percent of patients showed improvement in symptoms at 3 months but interestingly, some patients improved later during the first year after decompression. The results according to the presence or absence of syringomyelia are summarized in Table 2.

The flux MRI performed in 82.7% of the cases had made it possible to evaluate the effectiveness of the treatment by highlighting the presence of the CSF flow at the cranio-occipital level. Radiological monitoring of the size of the syrinx cavity revealed a disappearance in four cases (23.5%), a decrease in size in eight cases (47.1%) and stability in five cases (29.4%). When looking at the degree of clinical improvement in relation to radiological change of the syrinx volume, stabilization or disappearance of syrinx size was observed in 83% of patients with a good clinical outcome.

4. Discussion

4.1. Discussion of our findings and results

From the clinical point of view, stress or positional headaches, neck pain and instability generally respond well to surgical treatment. We consider that the surgical technique used allowed a clinical improvement or cure in 82.8% of our cases, which compares favorably with the data reported in the literature [4,5] and with a low rate of complications. These results are linked to a rigorous selection of eligible patients to surgery, a reproducible technique and a protocolized sequential follow-up of our patients. Another important point was the careful analysis of early failure. In these cases the contribution of multimodal MRI was paramount to identify a residual obstacle (adhesion of the inner surface of the plasty or arachnoiditis) that led to revision surgery which allowed a functional improvement (Fig. 2). When combined with syringomyelia, a neurological improvement can be clinically observed in 40 to 60% of the cases, among those with sensory and painful manifestations [2]. An analysis by Durham and Fjeld-Olenec [6] reported an improvement in the size of the syrinx by 50% with simple sub-occipital craniectomy and 80% to 100% if the craniectomy was associated with duraplasty. In our study, 83% of the patients with a decrease or disappearance of the syringomyelic

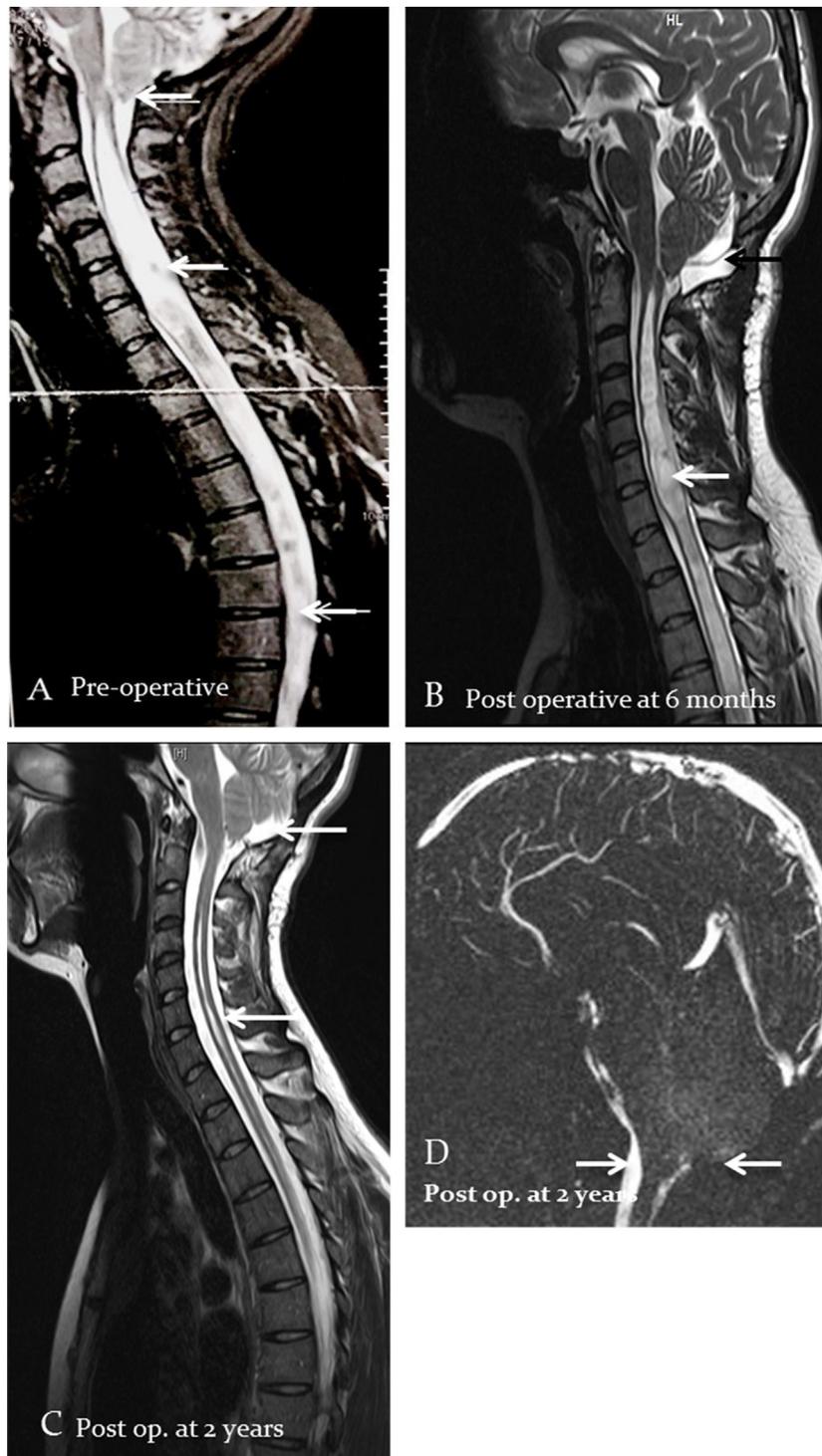


Fig. 1. A. Preoperative MRI in the T2 sagittal sequence showing a CM-I (white arrow) associated with pan medullary syringomyelia (white arrows). B. Postoperative MRI at 6 months after suboccipital decompression, C1 laminectomy and duraplasty in T2 sagittal sequence showing adhesion of duraplasty at the posterior cerebral fossa (black arrow) and a decrease in size of the syrinx cavity (white arrow). C. Postoperative 2 years follow-up. MRI in T2-imaging demonstrating an artificial cisterna magna, rounded shape of tonsil and reduction in syrinx size (white arrows). D. On the flow MRI sequence we note the presence of CSF flow around the foramen magnum (white arrows).

cavity had good clinical outcomes. This rate was quite similar for the ones who displayed a stable cyst which indicates that this radiological parameter does not influence the clinical outcome. This has been already published previously. Precise evaluation of clinical outcomes after surgical management of type 1 CM has been difficult to obtain at a large scale [7] (Table 3).

Several scores have been used in various studies to evaluate the clinical outcomes of patients with CM-I: Asgari score (modified)

[19,28], CCOS [29], JOA score [30], Klekamp & Scohier score [31,32], the Limonadi score [33] and the VAS-pain [34]. However, there is currently no generalized validated score in the literature that allows for a common and informative evaluation to improve patient care. Nevertheless, an effort to better assess our patients before and after surgery should be stressed since our goal is to refine the case selection and stratify our results according to the preoperative symptoms.

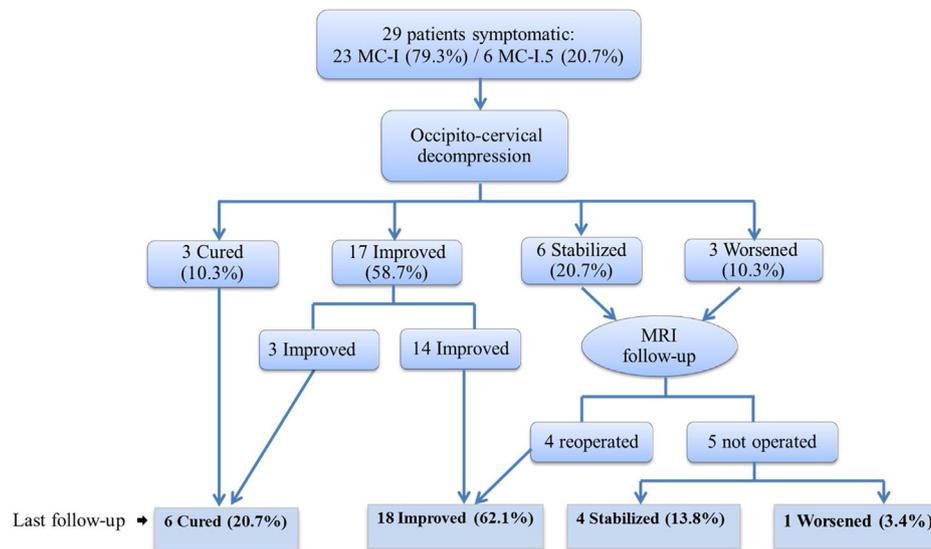


Fig. 2. Organizational chart summarizing the results of our patients during the last consultation.

Table 2
Global results which group the good results including patients cured and improved on the one hand and bad results comprising stabilized and aggravated patients on the other hand.

Results	Total (%)	With syrinx Number of cases (%)	Without syrinx Number of cases (%)
Good (Cured/improved)	24/29 (82.8%)	14/17 (82.3%)	10/12 (83.4%)
Bad (Stabilized/worsened)	5/29 (17.2%)	3/17 (17.6%)	2/12 (16.7%)

Table 3
Summary of clinical and radiological findings of the literature in association with CM and syringomyelia after decompression and/or shunt surgery.

Studies	Techniques	Type of study	Number	Clinical results (%)			Syrinx in post op. (%)		
				IM	ST	WO	IM	ST	WO
Vaquero 1990 [8]	ODD + OA	Monocentric	15	4 (27)	6 (40)	5 (33)	14 (93)	1 (7)	0
	SH		15	10 (67)	2 (13)	3 (20)	15 (100)	0	0
Pillay 1991 [9]	ODD + OA	Monocentric	17	9 (53)	6 (35)	2 (12)			
	Tognetti, 1993 [10]	Monocentric	17	14 (82)	2 (12)	1 (6)	12 (100)	0	0
Van Velthoven, 1993 [11]	SH		12	4 (33)	5 (42)	3 (25)	12 (100)	0	0
	ODD + OA	Monocentric	25	10 (40)	9 (36)	6 (24)			
Versari, 1993 [12]	ODD + OA	Monocentric	40	25 (63)	9 (22)	6 (15)	28 (70)	12 (30)	0
Fisher, 1995 [13]	ODD + OA + RT	Monocentric	16	9 (56)	7 (44)	0	14 (93)	1 (7)	0
Guyotat, 1998 [14]	ODD	Monocentric	50	18 (36)	14 (28)	18 (36)	14 (58)		
Aghakhani, 1999 [15]	ODD + OA	Multicentric	242	92 (38)	121 (50)	29 (12)	36 (15)		
	SH		31				(7)		
Munshi, 2000 [16]	ODD + OA	Monocentric	12	10 (84)	2 (16)	0	9 (100)	0	0
	OD		7	5 (71)	2 (29)	0	3 (50)	0	3 (50)
Sindou, 2001 [17]	ODD	Monocentric	15	12 (80)	3 (20)	0	9 (60)	6 (40)	0
Perrini, 2007 [18]	ODD	Monocentric	24	21 (87)	3 (13)	0	20 (83)	4 (17)	0
Koç, 2007 [19]	ODD + OA	Monocentric	18	11 (61)	5 (28)	2 (11)	13 (73)	5 (27)	0
Zhang, 2008 [20]	ODD + RT + SH	Multicentric	218	206 (94)			145 (67)	57 (26)	16 (7)
Chauvet, 2009 [21]	ODD	Monocentric	5	2 (40)	3 (60)	0	4 (80)	1 (20)	0
Aghakhani, 2009 [22]	ODD + OA	Monocentric	156	99 (63)	48 (31)	9 (6)	118 (76)	33 (21)	5 (3)
Lee, 2010 [23]	ODD	Monocentric	25	20 (80)	4 (16)	1 (4)	17 (68)	7 (28)	1 (4)
Spena, 2010 [24]	ODD	Monocentric	36	29 (80)	4 (11)	3 (8)	29 (80)	7 (20)	0
Aferi, 2012 [25]	ODD + RT	Monocentric	109	102 (93)	4 (4)	3 (3)	32 (29)	60 (55)	17 (16)
El Ghandour, 2012 [26]	ODD	Monocentric	12	4 (33)	8 (67)	0	9 (75)	3 (25)	0
	ODD + OA		14	8 (57)	6 (43)	0	13 (93)	1 (7)	0
Jian, 2016 [27]	ODD + SH		6	2 (33)	4 (67)	0	6 (100)	0	0
	OD	Multicentric	216	159 (74)	40 (18)	17 (8)	108 (77)	25 (18)	7 (5)
Our study, 2017	ODD		547	449 (82)	79 (15)	19 (3)	319 (87)	34 (10)	12 (3)
	ODD + RT + SH		187	169 (90)	14 (8)	4 (2)	91 (72)	28 (30)	7 (8)
	ODD + SH		82	49 (60)	8 (10)	25 (30)	31 (49)	12 (19)	21 (32)

ODD: osteo-dural decompression; OD: bone decompression; OA: opening of the arachnoid layer; DU: duraplasty; RT: resection of cerebellar tonsils; SH: shunt; IM: improved; ST: stabilized; WO: worsened.

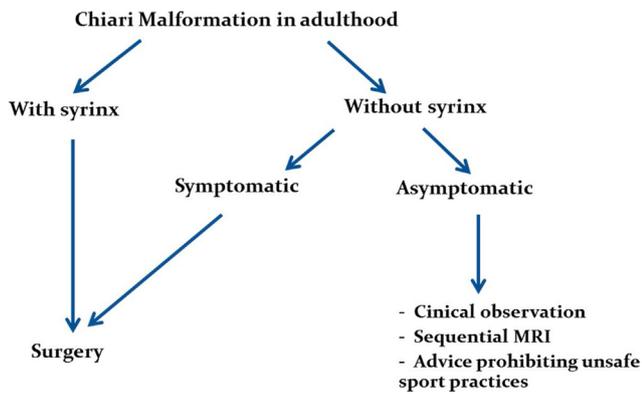


Fig. 3. Management of Chiari malformations in adulthood: decisional algorithm.

4.2. Selection of patients for surgery

In the literature, there is a consensus about the surgical treatment of symptomatic patients with CM-I, with syringomyelia [22] but surgery for patients with CM-I without syringomyelia remains a subject of debate [35]. The surgical indication must be carefully laid out after a lengthy observational period, as surgical complications are not uncommon [27]. The surgical treatment of CM is recommended when the malformation is symptomatic, provided that the present manifestations are indisputably related to the malformation or that the medullary MRI shows associated syringomyelia. Concomitant existence of CM and a syrinx is considered an absolute indication for decompression surgery, although some authors have reported cases of asymptomatic syrinx patients who have experienced a spontaneous disappearance of the syringomyelic cavity [27]. Malformations accidentally discovered by an MRI performed for various reasons may eventually become symptomatic. They should be monitored and prohibited from certain sport activities (see flowchart—proposal for surgical indications) (Fig. 3). When patients are considered eligible for surgery there is no justification to delay this treatment particularly if symptoms are due to syringomyelia where it has been showed that the neurological outcome was influenced by surgical timing [22].

4.3. Surgical technique

There are a variety of surgical approaches to treat CM in adults. Gardner had initially proposed to plug the obex using a muscle fragment [36], Rhoton recommended the direct opening of the syrinx, Menezes preconized a posterior fossa decompression with duraplasty and the fourth ventricle to subarachnoid shunt was made in all patients with CM type 1 with hydromyelia [37]. The contribution of these gestures to osteo-dural decompression was not validated while they brought their own risks. In a recent comprehensive study analyzing the full panel of techniques [27], osteo-dural decompression (ODD) with enlarged duraplasty showed more favorable results than other combined surgical methods in patients with CM1, with or without syrinx. Indeed, ODD offered better clinical results than simple bone decompression and provided more complications when paired with tonsillar resection. The cysto-arachnoidal shunt displayed the lowest rate of clinical improvement.

Osteo-dural decompression answers to the goals of surgery, allowing the creation of a new large cistern and thus restoring the flow of CSF in intracranial and intraspinal arachnoid spaces. However, there is no consensus about the extent of bone decompression and a large number of surgical adjuvants. Many neurosurgeons

recommend a large sub-occipital craniectomy with a wide resection of the foramen magnum which extends laterally to the two occipital condyles [22]. The collapse of the cerebellum can be observed in the case of a broad lateral craniectomy, which requires correction by cranioplasty [38]. Others adopt a limited occipital craniectomy to reduce the risk of postoperative cranio-cervical instability and cerebellar collapse [18]. In our study, the size of the flap is equal to 4×4 cm associated with the resection of the posterior arch of C1, which allows an appropriate restoration of the flux with minimal risk of caudal migration of the cerebellum. After opening, careful inspection of the intradural operative field under microscope looks useful to check for potential arachnoid partitioning or adhesions to the pia. However, intra-arachnoid extensive dissection should be avoided in order to limit the risk of bleeding and to avoid postoperative arachnoiditis which has been considered to be a source of failure [27]. Resection of the cerebellar tonsils may improve symptoms in adult patients with CM [39]. However, it has been discouraged in more recent publications due to the high morbidity rate [27]. In adult CMs associated with syringomyelia, some authors have recommended an intra-arachnoid exploration to expose and open the foramen of Magendi [26]. In our experience, the fact that symptoms improved equally regardless of the evolution of the syrinx volume does not give us additional arguments to move toward this surgical step.

The avoidance of adding a duraplasty certainly reduced the risk of postoperative complications such as acute hydrocephalus, aseptic meningitis, leakage of CSF, or pseudomeningocele [18] but the amount of dura that need to be duplicated in this technique is pretty difficult to assess. However, this duplication guarantees the expansion of the dura that will allow flow restoration. This may explain why the results of this strategy looks suboptimal [27], duraplasty with an autologous material or artificial substitutes is done with the aim of increasing the potential space for the nervous structures and promote the flow of CSF between the subarachnoid spaces intracranially and spinally [22]. The discussion about the optimal dural type of plasty continues [40,41]. The ideal duraplasty should allow for a good watertight closure without marked arachnoid adhesions nor causing an inflammatory response. In our opinion, tack-up sutures of the duraplasty to the bony boundaries and to the muscles at the center, could reduce the adhesions of its inner layer to the underlying arachnoid which was responsible for early failure in several of our patients.

4.4. Postoperative complications

The incidence of postoperative complications is influenced by the technique that is used. Aseptic meningitis and CSF leakage are the most frequent complications [27]. Meticulous watertight duraplasty with proper closure of the aponeurotic and cutaneous layers might reduce the incidence of incisional CSF leaks. Asymptomatic pseudo-meningocele is frequently discovered on the early postoperative MRI and does not interfere with the results as long as the collection is not compressive.

4.5. Limitations of our study

The reduced number of cases, the relatively short follow-up, lack of evaluation scores and the retrospective design of this study represent its principal limitations. Nevertheless, the study of our complications and the way we assessed and showed the course of the clinical evolution provided original information.

5. Conclusion

The indication for surgery of symptomatic CMI and CM1.5 adult patients is now well established. In this study we showed that

an occipital craniectomy of adequate size with resection of the posterior arch of C1, combined with an enlargement duraplasty was associated with more than 80% of good results with acceptable morbidity. This simple and reproducible surgical technique significantly improved the symptoms of paroxysmic intracranial hypertension and those attributed to syringomyelia. The majority of patients improved within 3 months and early failures could benefit from a revision surgery in cases of insufficient restoration of CSF flow.

Disclosure of interest

The authors declare that they have no competing interest.

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