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ORIGINAL ARTICLE

Surgical management of syringomyelia unrelated to Chiari malformation or spinal cord injury

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Abstract
Purpose Syringomyelia is a misleading disease since the problem always lies elsewhere. Arachnoiditis, because it is radiographically difficult to discern, is an especially insidious cause. To better guide selection from among surgical treatment options for syringomyelia, we reviewed our case series of patients without Chiari malformation or spinal injury.
Methods Excluding syringomyelia due to Chiari malformation, spinal cord injury, and tumors, 32 patients (mean age 44 years) were operated on between 1995 and 2013 and followed up for a mean of 53.8 months. Presumed causes at diagnosis, clinical and radiological findings, type of operation, clinical and radiological outcome were reviewed.
Results Duration of clinical history varied widely (range 6–164 months). Clinical assessment was based on the McCormick classification (15 independent, 17 dependent). Causes included birth trauma, pyogenic meningitis, tuberculous meningitis, postoperative scarring, dysraphism, and basilar impression. Treatment was local decompression with arachnoid lysis and shunts. Hindbrain-related syringomyelia was differentiated from non hindbrain-related syringomyelia. Hindbrain arachnoiditis was significantly associated with radiological findings at the foramen magnum ($p = 0.01$) and craniocervical decompression ($p < 0.03$), with good clinical and radiological outcome at 6 months and later follow-up controls ($p = 0.02$), whereas uneven results were observed in cases of non-hindbrain arachnoiditis.
Conclusions To remove the cause of syringomyelia, surgical planning will rely on thorough clinical history and accurate imaging to determine the site of cerebrospinal fluid obstruction. Craniocervical decompression to dissect basal arachnoiditis in the posterior fossa can be recommended in hindbrain syringomyelia. Treatment of non-hindbrain arachnoiditis is more controversial, probably owing to uncertainties about the extent of adhesions.

Keywords Syringomyelia · Surgical treatment · Arachnoiditis · Craniovertebral junction

Introduction

Syringomyelia is a polyetiologic progressive disorder characterized by longitudinal, intraspinal fluid-filled cyst formation [1–3]. Various different hypotheses have been advanced to explain its development in relation to obstruction of the spinal subarachnoid space. The prevailing theory is that disturbances in cerebrospinal fluid (CSF) flow in the subarachnoid space disrupt flow velocity, leading to passive distension of the spinal cord consequent to the Venturi effect (intramedullary pulse pressure theory) [4]. Two main categories of disturbances in CSF flow and related subarachnoid obstruction may be distinguished as: those due to dynamic obstruction and those due to fixed obstruction. Dynamic obstruction secondary to Chiari malformation results from the systolic motion of the

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cerebellar tonsils that increases propagation of the spinal CSF pulse wave to the distal CSF spaces (41–67 %) [5, 6]; fixed obstruction is typically seen in posttraumatic obstruction and is related to subarachnoid adhesions or vertebral fractures following spinal cord injury which limit the free propagation of the CSF pulse wave in the spinal canal, leading to transmission of the pulse pressure waves directly to the spinal cord (10–27 %) [6, 7]. Other causes of partial or complete obstruction of CSF flow include arachnoid scarring secondary to hemorrhage, infection, surgery, birth trauma, craniovertebral junction malformations, arachnoid cyst, and acquired or congenital spinal deformity (15–26 %) [6, 8]. The remaining cases are defined as "idiopathic syringomyelia" and refer to clinical cases of unknown origin without a syringomyelia-related history or underlying radiographic evidence (25–39 %) [6, 9]. Intramedullary or extramedullary tumor-related syringomyelia has been excluded from this count. Overall, this description fits with the Barnett classification [10].

Unlike Chiari malformation, in which surgical procedures and indications have been delineated, or post-traumatic syringomyelia, in which a clearer relationship between the acute event and the inflammatory response has been established, syringomyelia often remains a condition with an undefined pathological correlate, making it difficult for the treating physician to identify causal factors and subsequently deliver optimal patient care [2].

The aim of this retrospective study was to review the clinical history, radiological findings, and surgical procedures in syringomyelia patients without evidence of Chiari malformation or previous spinal cord injury to standardize a surgical approach as the best treatment option for specific pathological subgroups.

Materials and methods

Of the 122 patients operated on for syringomyelia at our hospital between 1995 and 2013, 32 (26.2 %) (13 males and 19 females; mean age, 44 years; range 20–67) were treated for syringomyelia unrelated to Chiari malformation, spinal cord injury or tumors. The clinical history and radiological findings of the different types of syringomyelia were reviewed. The presumed causes were: meningitis ($n = 12$, 5 of which due to tuberculous meningitis and 7 due to an unknown pathogen); craniocervical junction anomalies ($n = 9$); dystocic birth ($n = 4$); previous spinal surgery ($n = 2$); and unknown causes ($n = 9$).

Syrinx characteristics and CSF flow abnormalities were evaluated using myelography (MLG), magnetic resonance imaging (MRI) and cine MRI. MLG showed contrast medium block, irregular subarachnoid space, filling defects, sluggish contrast flow, and spinal cord atrophy. Additional imaging studies were sagittal and axial T1-weighted spin echo and T2-weighted fast spin echo MRI, phase-contrast cine MRI of CSF flow at the cisterna magna and spinal subarachnoid space. All images were reviewed to describe syrinx morphology and extension, meningeal thickening, subarachnoid scarring or spinal cord displacement (Fig. 1).

Radiological images of the craniocervical junction were reviewed for evidence of malformations. Characterization of radiographic abnormalities relies on lateral radiographs of the cervical spine with use of the Chamberlain line which extends from the posterior portion of the hard palate to the opisthion (posterior edge of the foramen magnum). The tip of the odontoid process typically lies just below the Chamberlain line; basilar impression is generally considered present if the extension is greater than 5 mm [11].

Notwithstanding accurate clinical and radiological data, the CSF level of obstruction is often not clear, i.e., whether the C1 level represents the end of the cyst or a level of obstruction. To better analyze the natural history of the cases and evaluate the surgical indications and approach, the patients were divided into two groups: group A ($n = 19$) comprised those with a syrinx extending from C1 to the conus ($n = 13$) and those with a syrinx extending from C1 to any level below before the conus ($n = 6$); group B ($n = 13$) comprised those with a syrinx extending from any cervical level below C1 to any level below ($n = 3$) and those with a syrinx at the thoracic level ($n = 10$).

Surgical indication was usually based on progressive neurological deterioration, since the duration of symptoms in this series ranged from 6 to 164 months; the primary endpoint was restoration of CSF flow. Surgical planning was based on clinical history and radiographic findings. Radiographic and clinical findings were compared to refine the final surgical strategy, especially the spinal level of surgery. In the absence of direct signs of CSF blockage at a clearly determinable level, the upper pole of the syrinx was the target.

The surgical procedures involved: posterior craniocervical decompression (CCD), which entails suboccipital craniectomy with opening of the foramen magnum and removal of the posterior arch of C1, lysis of arachnoid adhesions, restoration of communication between the fourth ventricle and the subarachnoid space, and expansive duraplasty; anterior craniocervical decompression (transoral approach) with C2 odontoidectomy and subsequent posterior occipito-cervical fixation (transoral approach); laminotomy or laminectomy, expansive duraplasty and lysis of arachnoid webs; laminotomy or laminectomy and placement of a syringo-subarachnoid shunt (SSS) or a syringo-peritoneal shunt (SPS).

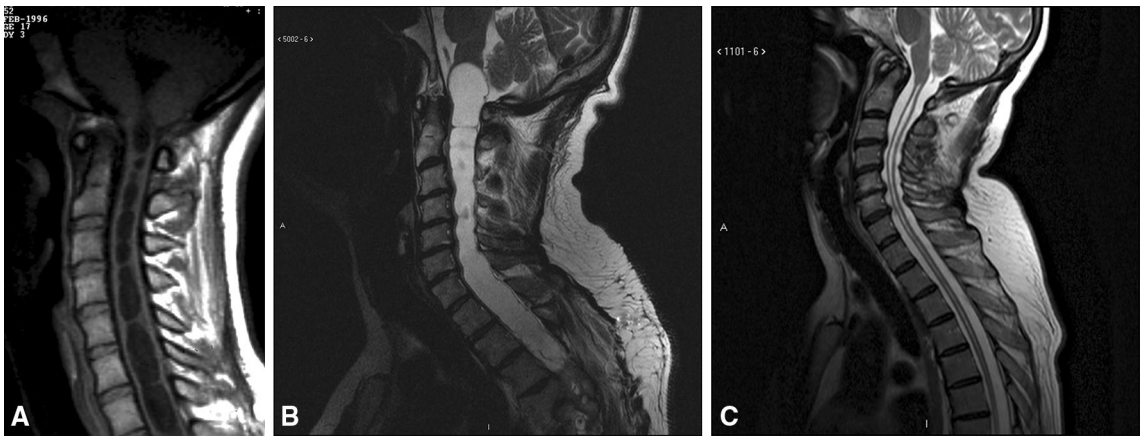

Fig. 1 Exemplifying features of syrinx morphology before treatment: "chambered" on a T1-weighted sagittal MR image (**a**), "convex" on a T2-weighted sagittal MR image (**b**), and "thin" on a T2-weighted sagittal MR image (**c**)

Outcome measurement at follow-up assessment was based on clinical and radiological evaluation. In order to standardize the data, the clinical outcomes were graded using the modified McCormick disability scale (grade I: intact neurologically, normal ambulation, minimal dysesthesia; grade II: mild motor or sensory deficit, functional independence; grade III: moderate deficit, limitation of function, independent with external aid; grade IV: severe motor or sensory deficit, limited function, dependent; grade V: paraplegia or quadriplegia, even with flickering movement) [12]. Data collection was performed on admission, at discharge, at 6 months and later follow-up controls. Clinical evaluation on admission and at follow-up was graded according to the modified McCormick scale, whereas at discharge, only a categorical description of clinical improvement/stability/worsening of preoperative symptoms was documented. Radiological assessment at 6 months and later was based on changes in syrinx morphology after surgery: complete shrinkage, incomplete shrinkage, no difference between preoperative and postoperative syrinx morphology. Clinical and radiological follow-up data were analyzed and compared to determine clinical and radiological outcome after surgery, which are not necessarily the same.

Results

There was no difference in the mean duration of clinical history between the two groups (74 vs. 82 months). The distribution of clinically related events was similar in both groups for the main cause, history of inflammatory disease, 6/19 (31 %) vs. 6/13 (46 %). Some other causes were site-specific: in group A dystocic birth ($n = 4$; 21 %), in group B myelitis ($n = 1$) and disc herniation ($n = 1$). The clinical features are presented in Tables 1 and 3. Table 2 reports the correlations tested using Fisher's exact test between preoperative, intraoperative, and postoperative findings for group A.

Group A: clinical and radiological outcome

Nineteen patients presented a syringomyelia with the upper pole at C1:13 were holocord syrinxes and 6 had a lower pole at the middle thoracic level. The functional status on admission was grade I ($n = 0$), grade II ($n = 2$), grade III ($n = 6$), grade IV ($n = 8$), and grade V ($n = 3$), showing that the majority of patients had severe impairment and were functionally dependent, irrespective of the extension of the syrinx.

Since single observations did not indicate the precise level of arachnoiditis (always present), we wanted to see whether correlations existed between history, clinical and radiological features (Table 2). Although the comparison between unknown and known history-related events and radiographic abnormalities did not appear significantly different, we further investigated the group, this time considering only the potentially undetermined conditions, including meningitis, which might have been diffuse, and unknown causes, while excluding focal causes, dystocic birth, and dysraphism. A history of inflammatory disease was associated with subarachnoid anomalies at the foramen magnum visible on radiographic images in 4 of 6 patients (66 %), 2 of which with subarachnoid scarring on MRI and 2 with CSF blockage on MLG. Conversely, in patients without an event related to syringomyelia, the radiological images revealed subarachnoid space anomalies in only 3 of 7 patients (42 %), thus demonstrating an association between CSF blockage on imaging, meningitis and arachnoiditis at the craniocervical level. However, MRI basal findings were weak signs and not convincing per se in all the five cases. In addition, the length of clinical history and

Table 1 Syringomyelia: group A

Clinical data						Radiological data			
No.	Age (years)	Sex	History-related events	Clinical history (months)	Admission (McCormick)	Syrinx length	Syrinx morphology	Level of abnormality	
								Intradural	Extradural
1	29	M	Unknown	6	II	Holocord	Thin	None	C1–BI
2	28	F	Meningitis	48	III	Holocord	Chambered	C1–MT–SS (MRI)	None
3	23	M	Dystocic	12	III	Holocord	Chambered/convex	C1–MT–SS (MRI)	C1–BI
4	33	F	Dystocic	136	III	Holocord	Chambered/convex	None	C1–BI
5	42	F	Unknown (recurrence)	36	III	Holocord	Convex	C1–CSF block (cMRI)	None
6	20	F	Dystocic	12	IV	Holocord	Chambered	C1–MT–SS (MRI)	C1–BI
7	49	M	Unknown	108	IV	Holocord	Chambered	C1–MT–SS (MRI)	None
8	66	F	Unknown	80	IV	Holocord	Thin	C1–MT–SS (MRI)	None
9	33	M	MM	104	IV	Holocord	Convex	None	None
10	38	F	Unknown	84	IV	Holocord	Chambered/convex	None	None
11	52	M	Unknown	100	IV	Holocord	Chambered	None	C1–BI
12	39	F	TM	72	V	Holocord	Chambered	CSF block-T7 (MLG)	None
13	33	F	TM (recurrence)	112	V	Holocord	Chambered	CSF block-T11 (MLG)	None
14	50	F	Unknown	24	II	C0–T5	Convex	None	None
15	66	F	Dystocic	120	III	C0–T8	Chambered	None	None
16	54	M	Meningitis (recurrence)	164	IV	C0–T4	Convex/thin	None	C1–BI
17	54	M	Meningitis	24	V	C0–T8	Convex	C1-C5-MT-SS (MRI)	None
18	33	F	Meningitis	84	III	C0–T6	Convex	None	None
19	44	F	Unknown (recurrence)	92	IV	C0–T8	Convex	None	None

Surgical data		Outcome at 6 months		Outcome at later follow-up		
Surgical approach	Operative notes	Radiological (shrinkage)	Clinical (McCormick)	Radiological (shrinkage)	Clinical (McCormick)	Time (months)
CCD	None	Complete	I	Complete	I	34
CCD+WL	Arachn	Incomplete	II	Complete	I	32
CCD+WL	Arachn	Complete	II	Complete	II	15
CCD+WL	Arachn	Incomplete	II	Complete	I	24
CCD+WL	Arachn	Incomplete	II	Complete	II	38
CCD+WL	Arachn	Incomplete	III	–	–	6
CCD+WL	Arachn	Incomplete	III	Complete	II	48
CCD+WL	Arachn	Incomplete	III	Incomplete	III	42
SSS	None	Incomplete	III	Incomplete	II	120
SSS	None	Incomplete	III	Incomplete (*)	IV	81
SSS	None	Complete	IV	Enlarged (*)	IV	84
SPS	Arachn	Incomplete	V	Incomplete	V	18
SSS	Arachn	Unchanged	V	Incomplete (*)	V	24
CCD+WL	Arachn	Complete	I	Complete	I	115

Table 1 continued

Surgical data		Outcome at 6 months		Outcome at later follow-up		
Surgical approach	Operative notes	Radiological (shrinkage)	Clinical (McCormick)	Radiological (shrinkage)	Clinical (McCormick)	Time (months)
CCD+WL	Arachn	Incomplete	III	Complete	III	25
CCD+WL	Arachn	Complete	III	Complete	III	72
Lam+WL	Arachn	Incomplete	V	Incomplete	IV	48
SSS	Arachn	Complete	II	Complete	II	61
SSS	None	Unchanged	IV	Incomplete (*)	IV	38

Dystocic dystocic birth, *MM* myelomeningocele, *TM* tuberculous meningitis, *MT–SS* meningeal thickening–subarachnoid scarring, *TDH* thoracic disc herniation, *MLG* myelography, *MRI* magnetic resonance imaging, *cMRI* cine MRI, *BI* basilar impression, *CCD+WL* craniocervical decompression+web lysis, *SSS* syringo-subarachnoid shunt, *SPS* syringo-peritoneal shunt, *Lam+WL* laminectomy+web lysis, *TOA* transoral approach, *CR+WL* cyst removal+web lysis, *Arachn* arachnoiditis, * patients submitted to new procedure, *C* cervical, *T* thoracic

Table 2 Correlation between clinico-radiological findings and treatment (group A)

	History-related events[a]	Clinical History (months)	Preoperative McCormick[b]	Syrinx morphology[c]	Radiological level of abnormality[d]	Arachnoiditis[e]	Surgical procedure[f]	Outcome at 6 months[g]	Outcome at later follow-up[h]
History-related events		–	–	–	–	–	–	–	–
Clinical history (months)			–	–	–	–	–	–	–
Preoperative McCormick				–	–	–	–	–	–
Syrinx morphology					–	–	–	–	–
Radiological level of abnormality						$p = 0.01$	–	–	–
Arachnoiditis							$p = 0.03$	–	–
Surgical procedure								$p = 0.02$	$p = 0.02$

[a] Comparison of unknown history-related events vs. all other related events

[b] McCormick grade 1–3 vs. 4–5

[c] Comparison of chambered morphology vs. other types of morphology

[d] Comparison of evident vs. not evident abnormality

[e] Comparison of evident vs. not evident arachnoiditis

[f] Comparison of CCD-WL vs. Shunt procedure

[g] Clinical outcome at 6 months

[h] Clinical outcome at later follow-up

the grade of functional impairment did not seem to significantly influence surgical approach or clinical outcome.

At 6-month follow-up assessment, clinical improvement in symptoms was observed in 10 of the 11 patients (91 %) who underwent CCD, but in only 3 of the 7 patients (42 %) who received a shunt ($p = 0.02$). This was clearly confirmed at later follow-up: 9 of the 10 patients submitted to CCD showed stable improvement as compared to preoperative clinical performance (1 patient lost to follow-up), whereas only 2 of 7 patients (26 %) who received a shunt showed stable improvement as compared to preoperative clinical performance ($p = 0.02$).

Radiological outcome showed complete syrinx shrinkage in 4 patients (37 %) submitted to CCD and incomplete shrinkage in 7 (63 %) (Figs. 2, 3); at later follow-up, complete syrinx collapse was seen in 5 of the 7 patients

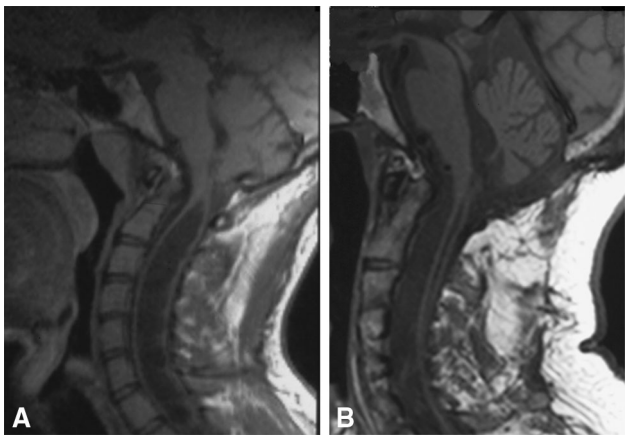

Fig. 2 T1-weighted sagittal MR image showing syringomyelia and craniocervical malformation with basilar impression (**a**). MR image showing marked shrinkage of the syrinx at one year after CCD (**b**)

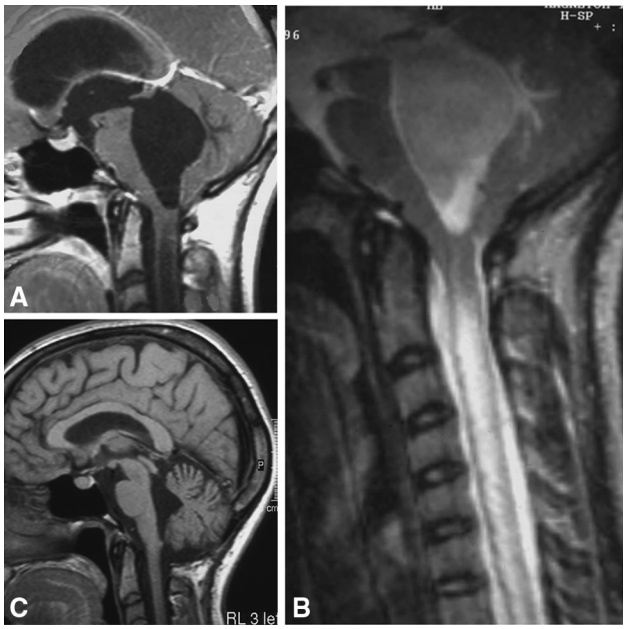

Fig. 3 T1-weighted sagittal MR image showing syringomyelia associated with hydrocephalus. Note the "tight cisterna magna" (**a**). T2-weighted sagittal MR image showing cervical extension of the syrinx (**b**). The last postoperative image shows no evidence of the syrinx and patent opening between the fourth ventricle and the subarachnoid space, as demonstrated by the normal size of the ventricular system, the restoration of central nervous system morphology, and the subarachnoid space (**c**)

(1 lost to follow-up) (Fig. 4). In 5 of the patients (71 %) who received a shunt, syrinx shrinkage was complete in 2, incomplete in 3, and unchanged in the remaining 2. At further follow-up, no changes in the syrinx were noted in 3 patients (42 %) (1 complete, 2 incomplete); 3 patients underwent a new SPS procedure without success. One patient with initially complete shrinkage relapsed and was not operated on.

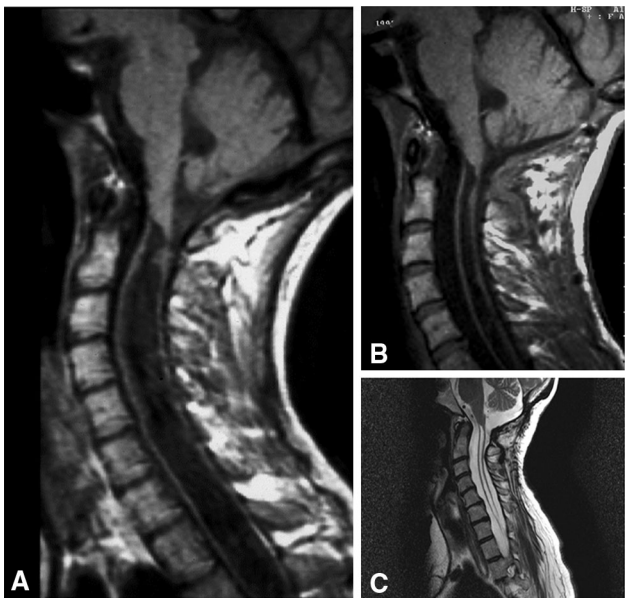

Fig. 4 T1-weighted sagittal MR image showing chambered syringomyelia with the upper level at C1 (**a**). The syrinx presents initial collapse at the first follow-up after 3 months (**b**). Two years later, further and definitive collapse of the syrinx can be seen (**c**)

The mean duration of follow-up was 48.6 months (range 6–120) and was fairly similar to that of group B (mean 56.9 months; range 18–128).

Group B: clinical and radiological outcome

History, clinical and radiological features, and outcome are reported in Table 3. In 10 of the 13 patients in this group the syringomyelia was thoracic and cervico-thoracic in 3. The functional status on admission was grade I ($n = 0$), grade II ($n = 1$), grade III ($n = 6$), grade IV ($n = 6$), grade V ($n = 0$), and not substantially different from group A. Differently from group A, however, combining radiological features and historic events scarcely helped the surgeon in operative planning. A history of inflammatory disease creating subarachnoid anomalies was radiographically visible in 3 of 6 patients (50 %) (1 with subarachnoid cyst on MRI and 2 with CSF blockage on MLG). Radiographic anomalies were visible in 3 of 5 patients (60 %) without clinically related past events (Fig. 5).

Clinical history of inflammatory processes carried a worse clinical outcome: in 3 of 6 patients (50 %) with a history of meningitis or other inflammatory process, clinical worsening of symptoms was observed at 6-month follow-up and continued at later follow-up controls. Only 1 of 6 patients (17 %) showed clinical improvement in symptoms at follow-up and 2 patients (33 %) showed no clinical changes after surgery. Clinical improvement in symptoms was noted in 3 of 5 patients (60 %) without a

Table 3 Syringomyelia group B

Clinical data						Radiological data			
No.	Age (years)	Sex	History related events	Clinical history (months)	Admission (McCormick)	Syrinx length	Syrinx morphology	Level of abnormality	
								Intradural	Extradural
1	65	F	Meningitis	84	IV	C3–L1	Convex	None	None
2	36	M	Unknown	116	II	C4–T4	Convex	None	C1–B.I.
3	67	M	Unknown	88	IV	C4–T4	Convex/thin	T1–T4–TM+AC (MRI)	None
4	46	F	Unknown	36	IV	T1–L1	Chambered/convex	None	None
5	56	F	TDH	72	III	T10–T11	Convex	T11/L1–AC (MLG)	None
6	54	F	Unknown	128	III	T11–L1	Convex	T11/L1–AC (MLG)	None
7	43	M	TM	92	III	T2–T9	Chambered	T8–T9–AC (MRI)	None
8	38	M	TM	54	IV	T2–L1	Chambered/convex	None	None
9	20	F	TM	120	III	T3–T8	Convex	None	None
10	44	M	TM	72	IV	T3–L1	Chambered	CSF block-T3 (MLG)	None
11	63	F	Myelitis	36	III	T6–T12	Irregular	CSF block-T8 (MLG)	None
12	55	F	MM	132	III	T7–T8	Convex	T6–T7–AC (MRI)	None
13	39	M	Unknown	36	IV	T7–T8	Thin	T6-CSF block (MLG)+T5–T6-AC (MRI)	None

Surgical data		Outcome at 6 months		Outcome at later follow-up		
Surgical approach	Operative notes	Radiological (shrinkage)	Clinical (McCormick)	Radiological (shrinkage)	Clinical (McCormick)	Time (months)
SSS	None	Unchanged	V	–	–	–
TOA	None	Complete	I	Complete	I	64
SSS	Arachn	Incomplete	IV	Incomplete	IV	18
SSS	None	Incomplete	IV	Enlarged	IV	36
SSS	Arachn	Incomplete	III	Incomplete	III	44
CR + WL	Arachn	Complete	II	Complete	II	28
WL	Arachn	Unchanged	II	Incomplete	II	35
SPS	None	Incomplete	III	Incomplete	IV	56
SSS	Arachn	Incomplete	III	Incomplete	III	18
WL	Arachn	Incomplete	V	Incomplete	V	78
WL + SPS	Arachn	Incomplete	IV	Incomplete	IV	84
CR + WL	Arachn	Incomplete	II	Incomplete	II	94
CR + SSS	Arachn	Unchanged	IV	Incomplete	III	128

MM myelomeningocele, *TM* tuberculous meningitis, *TDH* thoracic disc herniation, *MLG* myelography, *MRI* magnetic resonance imaging, *cMRI* cine MRI, *AC* arachnoid cyst, *BI* basilar impression, *CCD+WL* craniocervical decompression+web lysis, *SSS* syringo-subarachnoid shunt, *SPS* syringo-peritoneal shunt, *Lam+WL* laminectomy+web lysis, *TOA* transoral approach, *CR+WL* cyst removal+web lysis, *Arachn* arachnoiditis, *C* cervical, *T* thoracic, *L* lumbar

past clinical event clearly related to syringomyelia: in 2 cases there was no clinical change; no cases of clinical worsening occurred.

Symptom improvement was observed in 3 of 4 patients (75 %) submitted to lysis of arachnoid webs and worsening in 1; only 1 of 8 patients (12 %) submitted to a shunt procedure had a favorable clinical course, 2 worsened, and 5 were unchanged. This trend was confirmed at later follow-up, in which symptoms were unchanged.

On radiological outcome assessment at 6-month follow-up, 1 of the 4 patients submitted to lysis of arachnoid webs showed complete syrinx collapse, 2 showed incomplete collapse, and 1 showed no change in volume; at later follow-up, incomplete syrinx shrinkage was noted in the last case, and no further changes in the other 3 patients. Six of 8 patients (75 %) submitted to a shunt procedure showed incomplete syrinx collapse; no change in syrinx volume was noted in the other 2 patients; at later follow-up, the 6

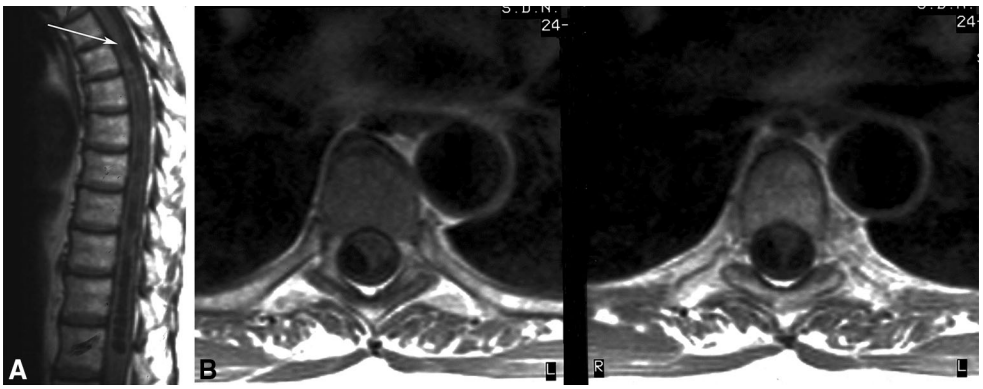

Fig. 5 T1-weighted sagittal MR image showing thoracic syringomyelia with a scalpel sign adjacent to the upper pole of the syrinx (**a**). At the same level on axial images (**b**), arachnoiditis causes severe constriction of the spinal cord which, together with the syrinx, should be considered a target of the operation

patients with incomplete syrinx collapse did not progress to complete syrinx shrinkage and 1 demonstrated syrinx enlargement. One of the last 2 patients was lost to follow-up; the other showed incomplete syrinx collapse at further follow-up.

At the last follow-up assessment, 90 % of patients improved after craniocervical decompression, 75 % after lysis of arachnoid adhesions of the spinal cord, and only 20 % of patients who received a shunt notwithstanding the higher radiological response (60 %).

Discussion

Although the incidence of idiopathic syringomyelia is decreasing, considerable uncertainty remains about the site and extension of CSF obstruction and the choice of treatment in many cases. While the cause of syringomyelia is unknown in about one-third of cases, many recognized causes have a complex mechanical or inflammatory pathogenesis which, albeit widely discussed in the literature, is difficult to interpret in individual cases [13–15]. And while the site of CSF blockage in Chiari malformation and spinal cord injury is clear, in other cases (arachnoiditis-related syringomyelia) diagnosis and treatment are less obvious [1, 4]. In this setting, we observed that the combined use of radiologic and history findings can assist the surgeon in deciding on the best surgical approach.

Dystocic birth, basilar invagination, disc herniation, dysraphism are all typical focal causes, whereas meningitis is more likely to be diffuse [16]. The treatment of arachnoiditis-related syringomyelia after meningitis is not always as ominous as expected but it will depend on syrinx configuration and radiological findings. We found that syringomyelia in the upper level at C1 indicates basal meningitis, which is focal in the vast majority of cases and can be successfully approached at the craniocervical level. In syringomyelia other than Chiari malformation, a similar but simpler pathogenesis can be considered as there is no tonsillar herniation or complicating mechanism of syrinx expansion. What remains are pathological changes in the subarachnoid space and the Magendie foramen, either of which may be the cause of syrinx formation [17]: after removal of the arachnoid adhesions, the CSF dynamics normalize, the syrinx size decreases, and symptomatic relief will follow [18].

Basal arachnoiditis was first described Appleby et al. [19] in 1969 and later reported in association with a number of causes: dystocic birth, tuberculous meningitis, pyogenic meningitis, basilar impression, and Chiari malformation [16, 20]. In 1990 Caplan et al. [21] classified it as diffuse and local, a distinct entity from spinal arachnoiditis, and in 1993 and in 1995 Milhorat et al. [22] better defined the characteristics describing a non-communicating and a communicating pathogenesis at this level, according to anatomopathological and imaging findings. Crowding of neural structures at the posterior cerebello-medullary junction could form an obstacle to CSF circulation, from both the fourth ventricle and the spinal canal to the subarachnoid space, a focal target for the operation. In our series, positive radiological findings were recognized at the same level in 68 % of cases. Interestingly, however, few supportive surgical experiences have been reported so far: in 1998 Iskandar et al. [23] observed resolution of different types of syringomyelia, by cause and extension, after posterior fossa decompression in 5 cases, and in 2002 Kyoshima et al. [24] resolved holocord syringomyelia by freeing the cisterna magna impacted by the tonsils (tight cisterna magna) in 4 cases of presumed idiopathic syringomyelia. Previously, only 22 cumulative cases have been traced, most of which with very favorable results [6, 19, 20, 25–27].

At the same level, diffuse arachnoiditis, extending also to the Luschka foramina, may lead to communicating syringomyelia, typically associated with hydrocephalus, which accounts for 10–15 % of hindbrain syringomyelia in historical series [17, 28, 29]. This view of a variable extent of hindbrain obstruction was clear to the pioneers, as Albouker [30] named basal arachnoiditis "foraminal syringomyelia", classified according to the number and degree of fourth ventricle foramen obstruction, but later often confused with spinal obstruction or idiopathic cases in clinical practice.

Despite improvements in MRI, choosing the surgical procedure may sometimes be problematic, in which instance elements from a thorough clinical history may reveal clues about the level of subarachnoid obstruction and its extent. It has also been demonstrated that even clear radiological findings often pass unrecognized initially or are overlooked, whereas subtle radiological findings (meningeal thickening or crowded posterior fossa) are properly interpreted only "a posteriori". In addition, radiological assessment may be inadequate and incomplete; ideally, it should encompass the brain and the spine in toto. This is especially true for spinal cord extremities distant from the syrinx: craniocervical malformations and occult spinal dysraphisms [16, 28].

In cases of previous meningitis or dystocic birth and intra- or extradural imaging abnormalities, we consistently found CSF blockage at the craniocervical junction where lysis of adhesions and osteodural decompression produced very good surgical results. While reserving this approach for cases of syringomyelia with the upper level at C1, we cannot exclude that, if other obstructions are absent, this procedure could also be effective in the treatment of syringes that do not reach this level [23]. The findings in the current series support the experience acquired with Chiari malformation, where the syringes were found at the lower border of the compressed segment of the spinal cord, separate from the fourth ventricle.

To date, the choice of appropriate treatment for arachnoiditis-related syringomyelia has been further complicated by changes in orientation of the literature over time. In the decade roughly from the 1980s to the late 1990s, many authors called attention to shunt procedures which had an effect on cyst filling. However, the transient effect and the scarce rate of clinical improvement downgraded drainage to second choice [31, 32]. Batzdorf approached the wide and neglected family of "primary spinal syringomyelia", classified as local or diffuse, by introducing the concept of surgical treatment of choice according to the same characteristics [3]. Local CSF obstruction should be approached with the firm intention to release it, while shunts are to be reserved for recurrences or diffuse spinal arachnoiditis. This principle was demonstrated effective by Klekamp et al. [13] in 1997 who performed only spinal procedures, shunt and decompression, most of which in patients with a history of spinal trauma. A high rate (83 %) of clinical stabilization was obtained with arachnoid dissection and decompression, provided that arachnoid scarring was limited. Minor experiences were reported by Lee et al. [18] in 2002 and by Nakamura et al. [33] in 2009, who reserved shunt placement for cases of diffuse arachnoiditis or with recurrences as palliative treatment on the basis of their short-term effect and the possibility to reoperate in case of failure [13]. Overall, improvement after shunt placement, regardless of modality and circumstances, syringo-subarachnoid or syringo-peritoneal or syringo-pleural shunts, primary operation or recurrence, accounts for 60 % of cases with a recurrence rate of 50 %, which is in line with our experience [13, 31, 32].

In our retrospective analysis of over 20 years, the prolonged mean clinical history underlines the dilemmas of which treatment is appropriate and whether one treatment can be reliably trusted over others in the single case, even though imaging is increasing helpful in identifying the level of abnormality. Sagittal and axial T2-weighted MR images can now be correlated with cardiac-gated 2D phase contrast, which is the method of choice to detect CSF flow abnormalities. Specifically, subtle arachnoid scarring may be indirect evidence for cord deformity or focal blurring of the syrinx wall, as well as direct evidence for an arachnoid web or cyst, obtained using 1.5T MR or 3T MR or 3D constructive interference in steady state (CISS) sequences, in order of power of resolution and sensibility [34–37]. CSF flow void signs over the cardiac cycle (cine MR) give a dynamic picture showing a short period of cranial flow (diastolic phase) followed by a sustained period of caudal flow (systolic phase), which can be altered by CSF obstruction, being of particular significance at the craniovertebral junction [38, 39]. However, direct intradural and dural decompression is hardly applicable in primary spinal syringomyelia, where the site and the extension of CSF obstruction are less clear than at the craniocervical level [18, 33, 40]. In addition, lysis of adhesions and shunting has not been described in any great detail and opinions on their use vary among authors. Accordingly, surgical planning can scarcely guide the surgical procedure and, conversely, the surgical procedure cannot confirm preoperative assessment [13, 41, 42]. In a time of rapidly evolving technological changes, more experience in such subgroups of rare diseases is needed.

Finally, arachnoiditis and spinal cord pulsation and dilatation all play an important role in the pathogenesis of clinical impairment [19, 40]. Imaging clearly shows the degree of cord deformity and constriction proximal to the syrinx, which is an additional factor in inflammatory-driven ischemia in determining spinal damage [37]. This role can be indirectly assumed as one of the explanations for

even short-term poor clinical outcome of drainage which collapses the syrinx while leaving the cord constriction untouched [18, 31].

In conclusion, notwithstanding the fundamental contributions by Williams and Milhorat in the early 1990s, non-communicating hindbrain obstructions other than Chiari malformation are overlooked and often confused with primary spinal syringomyelia [17, 31, 43]. Overall, these preliminary findings influence outcome in two ways: recognizing effective treatment to restore CSF circulation in patients with hindbrain syringomyelia due to basal arachnoiditis, and confirming that shunt procedures and arachnoid lysis have some limitations in remaining patients.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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