Syringomyelia

9

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Abstract

Syringomyelia is not a disease in its own right but a manifestation of another disease process, which incorporates either an obstruction of cerebrospinal fluid (CSF) flow in the spinal canal, tethering of the spinal cord, or an intramedullary tumor. Whenever a syrinx is demonstrated, clinical examination, analysis of the patient's history, and neuroradiological imaging have to identify the underlying cause of the syrinx. If this cause can be identified and treated successfully, the syrinx will regress, and clinical symptoms will improve or remain stable in the future.

The significance of spinal arachnopathies for development and treatment of syringomyelia in patients without a craniospinal malformation, a spinal dysraphic malformation, or an intramedullary tumor is still not widely recognized. This chapter describes diagnostic and management algorithms for spinal arachnopathies leading to syringomyelia with a special emphasis on posttraumatic syringomyelia as well as long-term results for these patients.

9.1 Introduction

The term syringomyelia was introduced by Ollivier d'Angers in 1827 [35] for cystic cavitations of the spinal cord. Syringomyelia describes a progressive accumulation of fluid inside the spinal cord. Up to this day, no pathophysiological concept for the development of syringomyelia is generally accepted. However, with the introduction of modern imaging techniques in the 1970s and 1980s, it became clear that a syrinx is always associated with additional pathologies in the spinal canal or

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craniocervical junction. This observation has changed treatment concepts for these patients in a fundamental way. If the associated pathology can be treated successfully, no further measures for the syrinx are needed. It is now widely accepted that syringomyelia is related to intramedullary tumors or pathologies that cause a disturbance of cerebrospinal fluid (CSF) flow or spinal cord tethering, i.e., fixation of the cord to the spinal dura by a thick filum terminale or other dysraphic lesions [17]. Table 9.1 gives an overview on the different pathologies related to syringomyelia in the author's series. About 74.5 % of patients with a Chiari I malformation developed a syrinx. Except for foramen magnum arachnoiditis, no other pathology causes syringomyelia in such a high proportion (Table 9.1).

Currently, syringomyelia is considered as an accumulation of extracellular fluid of the spinal cord [11, 17]. In case of intramedullary tumors, it is generally believed that alterations of the blood-spinal cord barrier play a major role [31]. But this may not be the only mechanism. It is noteworthy that infiltrating intramedullary tumors rarely produce syringomyelia, whereas a syrinx is a common feature of displacing neoplasms [24]. More information is available on the effects of CSF flow obstructions on the spinal cord from animal [27] as well as computer models [3]. Pressure changes in the subarachnoid space related to CSF flow obstructions may alter the distribution of extracellular fluid inside the spinal cord [27] which may then lead to syringomyelia [11, 17, 26]. Increased flow in the perivascular spaces has been implicated for this effect [2, 4, 17, 27, 43, 44]. If flow capacities in the extracellular space are exceeded, there appears to be an evolution from spinal cord edema, i.e., the so-called presyrinx state, to syringomyelia [8]. Intramedullary neoplasms and cord tethering may alter extracellular fluid movements to similar effects. Once syringomyelia has developed, the increased intramedullary pressure [32] and fluid movements inside the syrinx [1, 46] may lead to spinal cord damage [10, 39, 42] and progressive neurological symptoms.

Table 9.1 Pathologies associated with syringomyelia

Diagnosis	Total	Syringomyelia
Craniocervical junction	*756*	*556*
Chiari I	651	485 (74.5 %)
Chiari II	45	29 (64.4 %)
Foramen magnum arachnoiditis	33	33 (100 %)
Posterior fossa tumors	12	2 (16.7 %)
Posterior fossa arachnoid cysts	11	5 (45.5 %)
Spinal canal	*2143*	*853*
Posttraumatic arachnopathies	150	150
Nontraumatic arachnopathies	333	333
Intramedullary tumors	345	143 (41.4 %)
Extramedullary tumors	608	88 (14.5 %)
Extradural tumors	469	12 (2.6 %)
Tethered cord syndromes	170	59 (3.5 %)
Degenerative disc disease	68	68

9.2 Diagnosis

Intramedullary tumors associated with syringomyelia always display contrast enhancement on MRI. The most common entities are ependymomas (Fig. 9.1) and angioblastomas (Fig. 9.2). Whereas ependymomas associated with syringomyelia are solid space-occupying tumors, which are easily detected on MRI after application of gadolinium (Fig. 9.1), the diagnosis of an angioblastoma may not be straightforward. They tend to be localized in the dorsal root entry zone of the cord and may be quite small [24] (Fig. 9.2).

If tethered cord syndromes cause syringomyelia, these syrinx cavities tend to be rather small and of no clinical significance (Fig. 9.3). A large syrinx of the entire cord is rarely observed in patients with a tethered cord syndrome. The diagnostic challenge in these patients is the identification of all tethering elements, which tend to be localized below the lower pole of the syrinx. Such elements, which can be found in various combinations in a particular patient, are a thickened filum terminale, a split cord malformation (Fig. 9.3), or a lipoma fixed at the dura to mention the most frequent [19].

In patients with Chiari malformations, CSF flow can be compromised by cerebellar tonsils filling the space of the cisterna magna, by arachnoid scarring in the foramen magnum area, and by obstruction of the foramen of Magendie (Fig. 9.4). Whereas the obstruction by cerebellar tonsils is clearly apparent on a preoperative

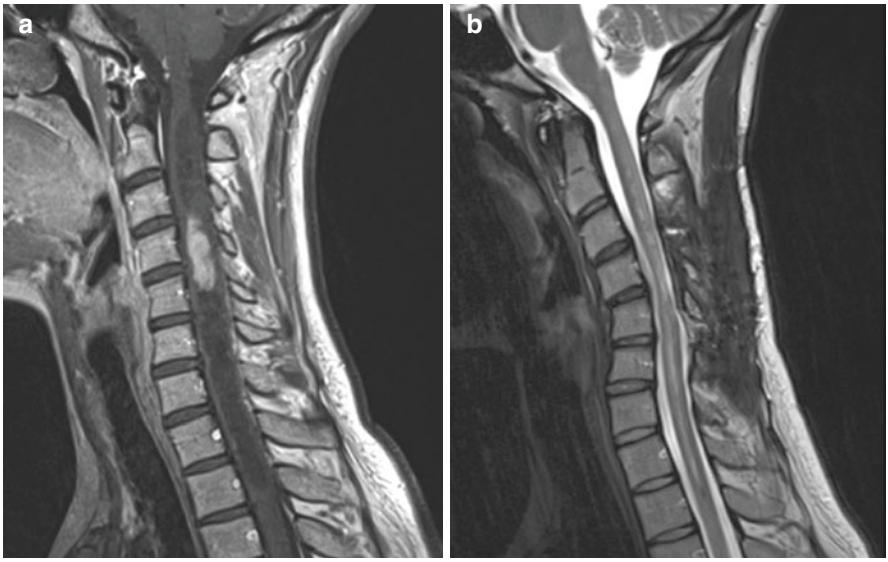

Fig. 9.1 (**a**) This T1-weighted MRI with gadolinium demonstrates an ependymoma at C4–C5 with associated syringomyelia above and below the tumor in a 26-year-old woman. The tumor shows bright contrast enhancement and is well demarcated from the surrounding cord tissue. (**b**) The postoperative scan shows the complete collapse of the syrinx after tumor removal. Despite reinsertion of the laminae, a kyphotic deformity developed

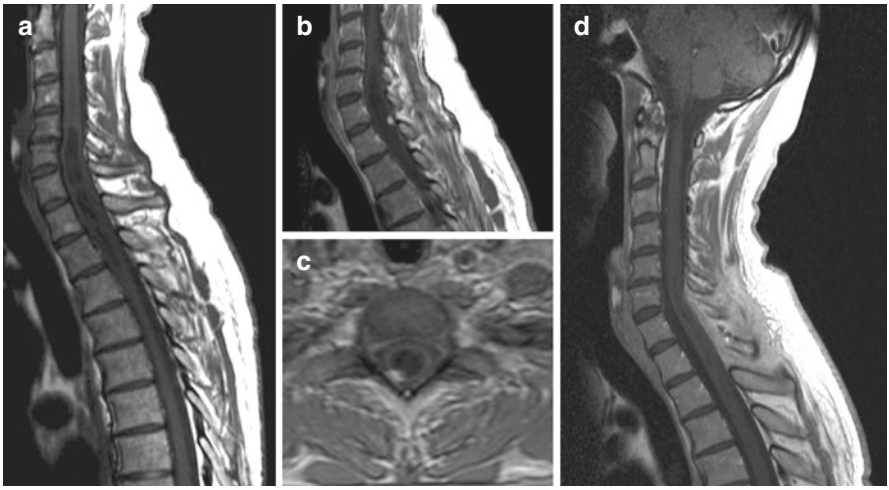

Fig. 9.2 (**a**) This T1-weighted MRI image of a 56-year-old man shows a syrinx between C5 and Th3. (**b**) After gadolinium application a small angioblastoma appears posteriorly at C7/Th1. The corresponding axial scan demonstrates the typical location of this tumor in the posterior root entry zone on the right side (**c**). (**d**) With removal of this tumor, the syrinx collapsed

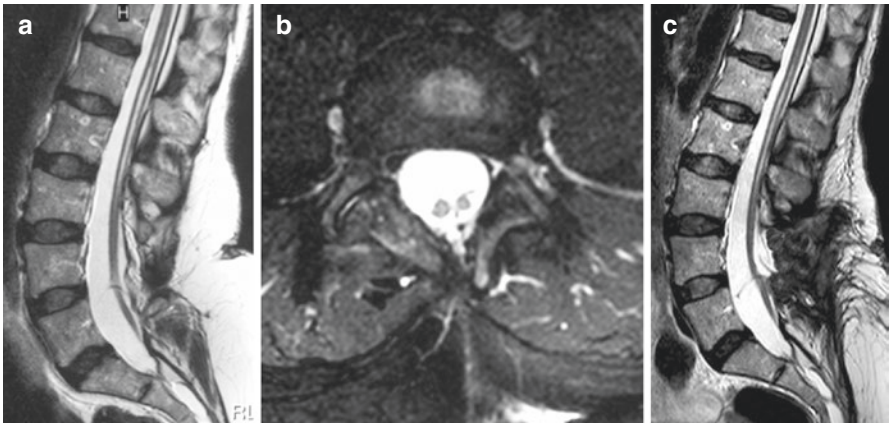

Fig. 9.3 (**a**) This T2-weighted image of a 53-year-old woman demonstrates the conus position at S1 and a small syrinx in the lower part of the cord, which exerts no space-occupying effect. (**b**) The axial scan at L4/5 shows a split cord malformation type 2 with a fibrous band splitting and fixing the cord at this level. Extradurally, this band was connected to a dermal sinus. Surgery required section of the filum terminale below S1 and untethering at L4/5. (**c**) The postoperative T2-weighted image shows no significant change of the syrinx despite complete untethering of the cord

MRI, arachnoid adhesions and an obstruction of the foramen of Magendie are mostly intraoperative findings and need special attention during surgery [18].

In posttraumatic syringomyelia, CSF flow obstruction may be caused by arachnoid scarring at the trauma level as well as narrowing of the spinal canal due to

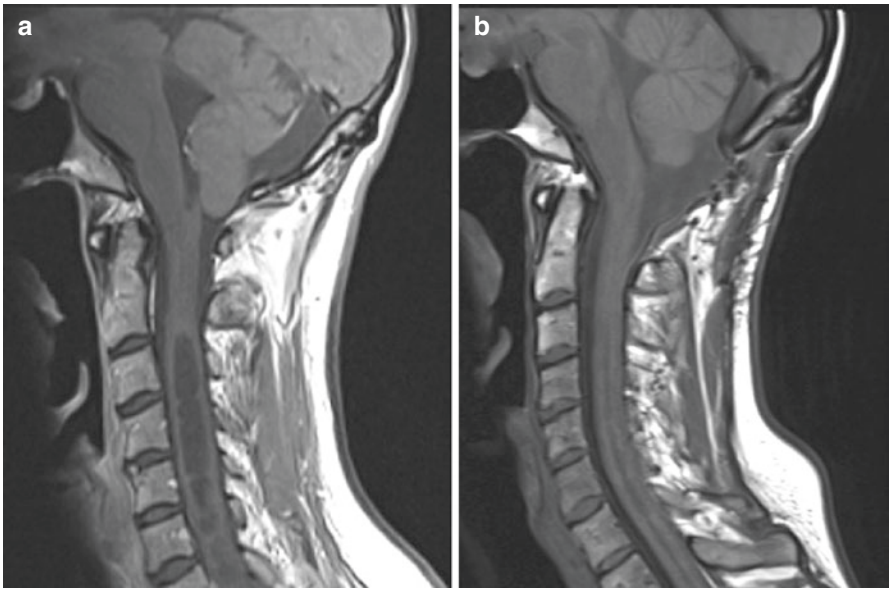

Fig. 9.4 (**a**) The T1-weighted MRI scan of a 47-year-old woman with a Chiari I malformation and a syrinx C3–Th2. (**b**) After decompression of the foramen magnum with a duraplasty, the CSF passage is free, and the syrinx has decreased

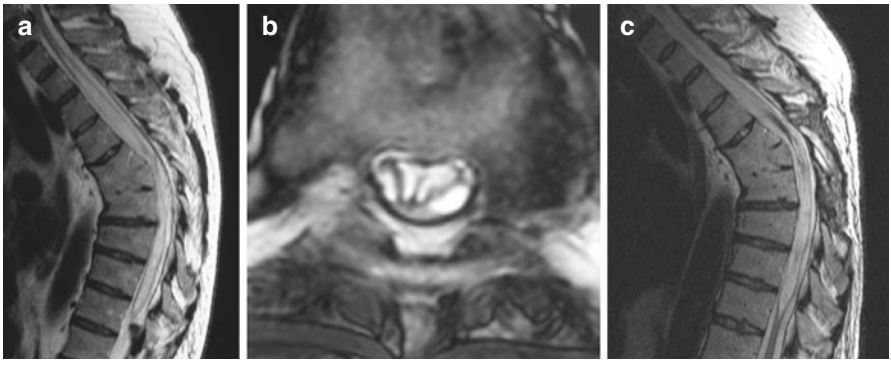

Fig. 9.5 (**a**) This T2-weighted MRI scan of a 49-year-old man shows a posttraumatic syrinx Th1–Th10 10 years after suffering a fracture at Th10 without any neurological deficits. A kyphotic deformity developed. (**b**) The axial scan at Th6 shows a posterior adhesion of the spinal cord to the dura resembling a posttraumatic arachnopathy. (**c**) The postoperative scan after arachnolysis and duraplasty at Th6 demonstrates a decrease of the syrinx (see also Fig. 9.8)

posttraumatic stenosis or kyphosis (Fig. 9.5). Furthermore, posttraumatic cord tethering may contribute to syrinx development. Depending on associated injuries to vertebral bodies and joints, MRI may not be sufficient for surgical planning. CT scans in bone window technique may be very helpful to localize and determine bony

landmarks for intraoperative orientation as well as the position of implants in patients who require a spondylodesis [21].

In the absence of a craniocervical malformation, an intramedullary tumor, or a history of spinal trauma, syringomyelia is still considered idiopathic by many physicians. However, these patients have to be evaluated very carefully for radiological and clinical signs of arachnoid pathologies in the spinal canal causing CSF flow obstructions [22] (Fig. 9.6). The syrinx starts at the level of obstruction and expands from there. If the syrinx expands in rostral direction, the obstruction will be found at the caudal end of the syrinx and vice versa. This also implies that the obstruction will most likely be found close to the largest diameter of the syrinx [22] (Fig. 9.6).

Due to the pulsatile movements of arachnoid septations, webs, or cysts, standard MRIs may not always be able to demonstrate an arachnopathy directly. With a

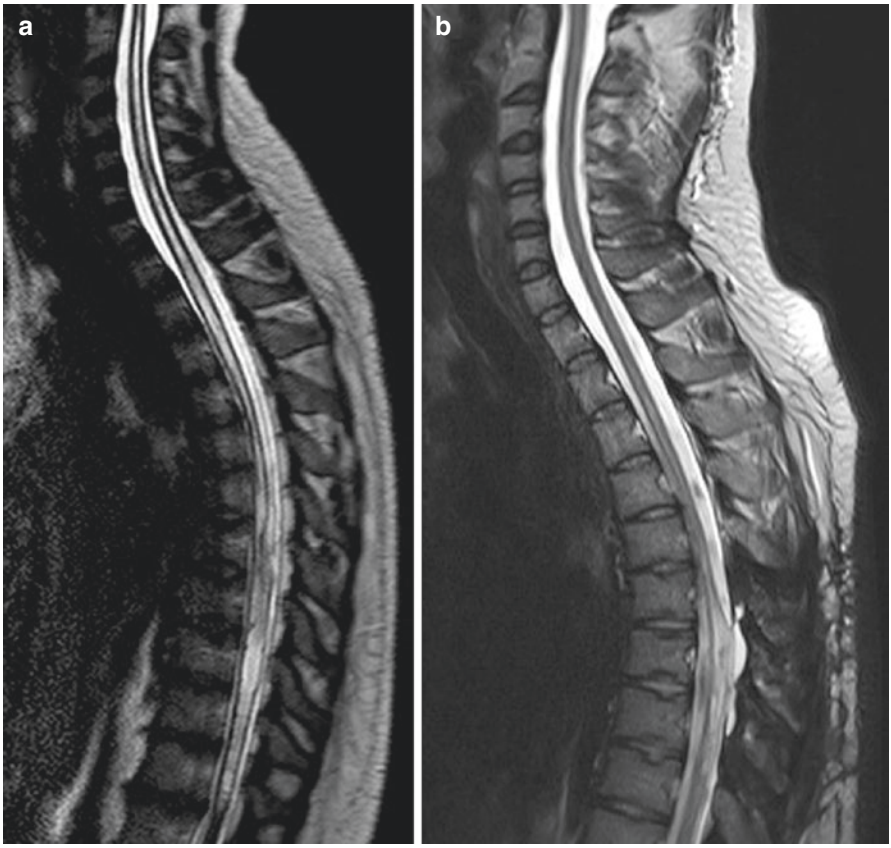

Fig. 9.6 (**a**) T2-weighted MRI scan of a 29-year-old man with a syrinx C2–Th11 and no history of trauma. At Th7 the cord appears compressed posteriorly and the syrinx caliber changes abruptly. At surgery, a circumscribed arachnopathy was evident at this level obstructing CSF flow and compressing the cord. (**b**) After arachnolysis and duraplasty at Th7, CSF flow at this level was established and the syrinx decreased

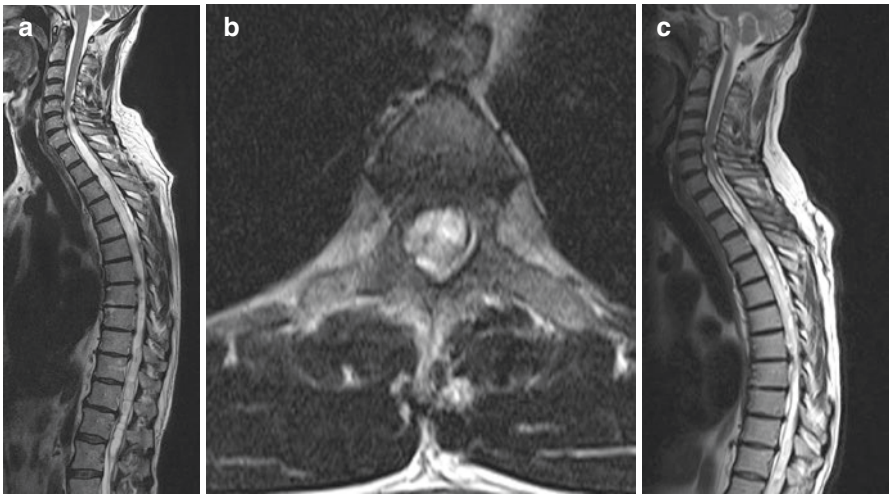

Fig. 9.7 (**a**) T2-weighted MRI scan of a 66-year-old man 5 years after suffering from a listeria meningitis. After recovery from this inflammation, the patient experienced a progressive paraparesis and ascending neurological deficits threatening his hand functions. The syrinx had reached up to C6. (**b**) The meningitis had led to severe arachnoid scarring around the entire cord from Th3 to Th10 as shown on the axial scan. (**c**) After placement of a thecoperitoneal shunt at the cervicothoracic junction lowering the subarachnoid pressure above the arachnopathy, the cervical and upper thoracic part of the syrinx has decreased as shown on this MRI after 6 months

history of spinal meningitis or subarachnoid hemorrhage [5], the often quite extensive arachnopathy is rather easy to diagnose by MRI (Fig. 9.7). Many arachnopathies, however, are quite discrete and extend over a few millimeters only (Fig. 9.6). Cardiac-gated cine MRI should be employed for such instances to study spinal CSF flow to identify areas of flow obstruction which may correspond to such circumscribed arachnoid pathologies [1, 22]. Sometimes significant flow signals can also be detected in the syrinx itself. In such cases, the highest flow velocities in the syrinx can be expected adjacent to the arachnoid scarring. The spinal cord should be studied with thin axial slices in T2-weighted images over the entire extent of the syrinx to search for areas of cord compression, displacement, or adhesion to the dura [7, 14, 22]. In the sagittal plane, the contour of the cord may appear distorted in areas of arachnoid scarring (Fig. 9.6). CISS (constructive interference in steady state) sequences can be used not only for the demonstration of the syrinx [12] but may also be helpful to detect arachnoid webs, scars, and cysts, because this technique is less susceptible to CSF flow artifacts [12]. Myelography and postmyelographic computer tomography (CT) are alternative methods to demonstrate arachnoid pathologies but have a lower sensitivity [22].

The sequence of events leading to a syrinx has implications not only for the neuroradiological appearance as just described but also for the evolution of clinical symptoms. The syrinx develops as a consequence of a series of events that originate from a pathology leading to CSF flow obstruction. For almost all entities leading to syrinx development, this process requires many years. Therefore, the first neurological

symptoms in the patient's history are commonly caused by this underlying pathology rather than the syrinx. In other words, a carefully taken clinical history can provide clues to the underlying pathology. If neurological signs spread to other parts of the body in an ascending pattern, the cause of the syrinx will be located at the lower pole of the syrinx and vice versa similar to the radiological evolution of the syrinx. Apart from trauma, arachnoid scarring may be related to infection [16] (Fig. 9.7), hemorrhage [5], irritation by outdated contrast agents such as pantopaque [28], or surgery, to mention a few [22].

It is always puzzling that patients may harbor a huge syrinx and yet have just minor symptoms with exactly the opposite observation for some smaller syrinx cavities associated with major neurological deficits. One explanation for this paradox may be that a great deal of the clinical problems are related to the underlying disease process causing the syrinx rather than to the syrinx itself [25]. This is particularly evident for syringomyelia associated with intramedullary tumors or cord tethering.

The classical symptom of syringomyelia is a dissociated sensory loss with loss of sensation for temperature and pain but preserved sensation for light touch. Pain related to syringomyelia is either permanent or aggravated by maneuvers such as coughing and sneezing and perceived in dermatomes corresponding to the level and extension of the syrinx. Syrinx pain represents neuropathic pain. In order to produce neuropathic pain, a syrinx must have reached the region of the dorsal horns close to the dorsal root entry zone. Quite commonly, patients report that their pain was associated with coughing or sneezing initially before becoming permanent. Other types of pain which patients report quite often, such as pain associated with exertion, cannot be explained by syringomyelia and are caused by other mechanisms such as muscular spanning to mention a common example. Late symptoms of syringomyelia are muscle atrophies corresponding to damage of ventral horn cells or trophic changes leading to skin and joint damages, particularly in the shoulder and elbow [25].

9.3 Management

Successful treatment of syringomyelia requires to treat the underlying cause – preferably before significant neurological deficits have developed. For syringomyelia associated with intramedullary tumors and Chiari I malformation, the rates for postoperative syrinx resolution are above 80 %, provided the tumor is removed and all components contributing to CSF flow obstruction in Chiari malformations have been surgically addressed, respectively [18, 20]. Syrinx cavities related to tethered cord syndromes tend to be of small caliber and extension. Therefore, complete untethering caused a postoperative reduction in just 13 %, whereas the syrinx remained unchanged in 87 % [19].

Successful treatment of syringomyelia related to spinal rather than craniocervical CSF flow obstructions is much more challenging. The underlying causes are more difficult to identify and to deal with surgically. For this reason, surgery on spinal arachnopathies for treatment of syringomyelia should be reserved for patients with progressive symptoms. Nevertheless, treating the cause of the syrinx with arachnolysis, untethering the cord, and duraplasty is rewarded by considerably better results compared to syrinx shunting procedures [21–23, 25, 33, 37].

Neuropathic pain and dysesthesias, particularly those of burning character, may be major clinical problems. Even though these may improve to some degree with successful treatment of the syrinx, this is never certain. Therefore, the decision for or against surgery should be based on the course of neurological signs and symptoms rather than pain syndromes alone [22].

In general, surgery can be recommended for patients with arachnoid scarring limited to about 2–3 spinal segments [22] (Fig. 9.5). All operations are performed in prone position. Laminotomies are recommended to reinsert the lamina at the end of the operation with titanium miniplates. After exposure of the dura, the extent of the arachnoid pathology and the syrinx can be visualized with ultrasound. Pulsations of syrinx fluid and CSF may become visible. Most importantly, the safest spot for opening of the dura can be chosen with this technique. As contamination of the CSF with blood may cause inflammatory reactions of the arachnoid, great care must be taken to achieve good hemostasis. For this purpose, the entire surgical field is covered by moist cottonoids, which keep soft tissues moist and soak up any minor bleeding. Then the dura is opened under the operating microscope in the midline without opening of the arachnoid. Once the dura is held open with sutures, the arachnoid pathology can be studied, and adequate exposure cranially and caudally is ensured in order to gain access to a normal and unaffected subarachnoid space on either end (Fig. 9.8a). Obviously, any surgeon should be familiar with the normal anatomy of the spinal subarachnoid space [34]. The posterior subarachnoid space is divided in two halves by a posterior longitudinal arachnoid septum. This septum extends between the outer arachnoid layer and an intermediate layer on the cord surface. The insertion on the cord surface is related to the midline dorsal vein. Further strands of arachnoid may be encountered in the posterior and lateral subarachnoid space. These arachnoid webs and septations are a physiological feature of the thoracic spinal canal. In the cervical area, the posterior longitudinal septum may be absent, and lateral arachnoid webs between nerve roots, which are considered to promote CSF flow in the thoracic spine, are not present. Under normal conditions, no arachnoid webs or septations exist in the entire anterior spinal subarachnoid space. A good landmark for dissection are the dentate ligaments, which originate from the spinal cord pia mater, run between posterior and anterior nerve roots, and insert close to the dural nerve root sleeve. With a microdissector, arachnoid and dura can be separated from each other without any problem in areas without arachnoid scarring, i.e., at either end of the exposure. In the area of scarring, sharp dissection with microscissors is usually required to achieve this (Figs. 9.8b, c). At the level of CSF flow obstruction, the arachnoid may become densely adherent to the cord surface. With opening of the rostral and caudal subarachnoid space, CSF flushes into the surgical field, and often the cord, which was distended by the syrinx, starts to pulsate, and the syrinx may collapse at this point. The arachnoid scar can be resected layer by layer leaving a last sheath on the cord surface to avoid injury to the cord or surface vessels (Figs. 9.8d, e). This last layer resembles the intermediate arachnoidal layer mentioned above. In this way, a free CSF passage in the posterior subarachnoid space can be created in every patient across the region of the arachnopathy. Dissection is then continued laterally on either side toward the dentate ligaments. This leads to a complete untethering of the cord in the majority of cases. No arachnoid dissection should be performed anteriorly of the dentate

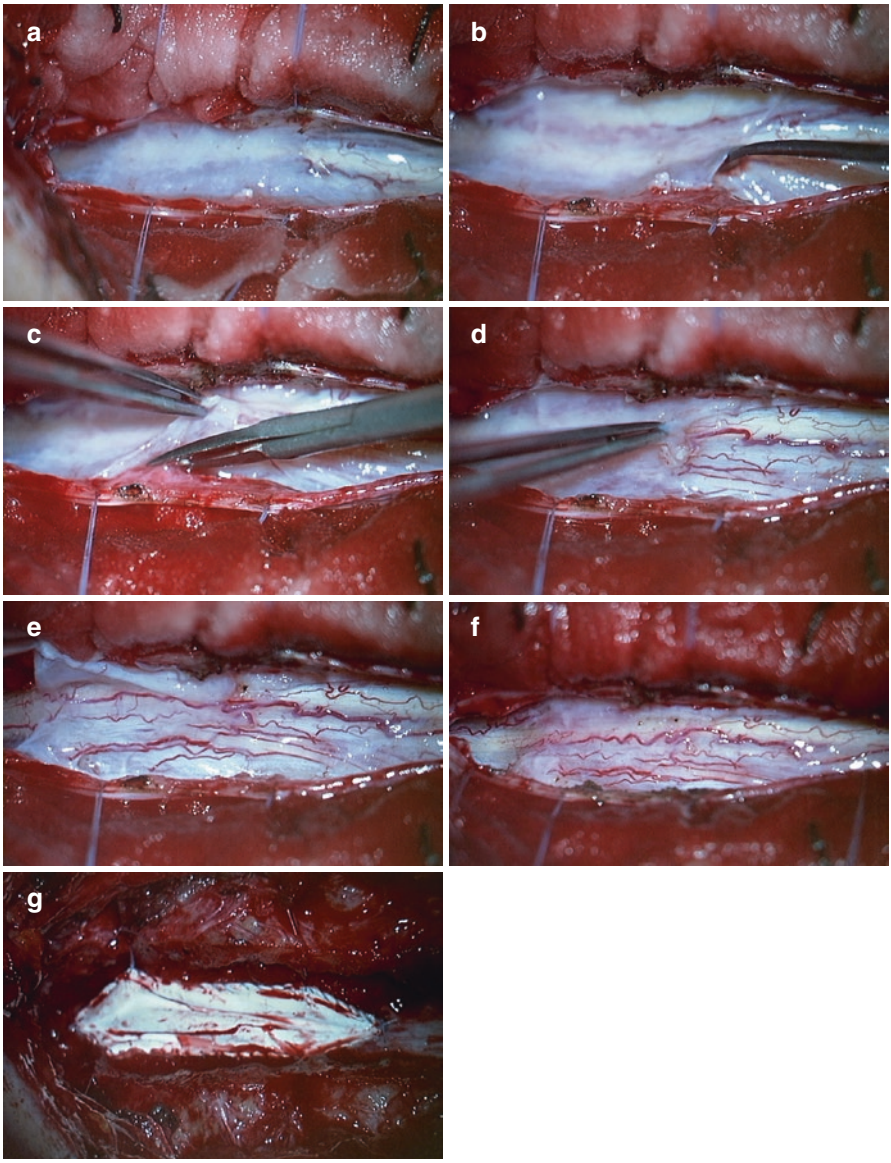

Fig. 9.8 (**a**) This intraoperative view after dura opening shows the posttraumatic arachnopathy at Th6 corresponding to Fig. 9.5. The arachnoid appears translucent in the right half of the exposure marking the lower end of the pathology. (**b**) The arachnoid is adherent to the dura requiring sharp dissection to separate it (**c**). Starting below the arachnopathy, the arachnoid is incised and dissected off the spinal cord. The arachnopathy is gently held under slight tension with microforceps (**d**) and cut preserving the blood vessels on the cord surface which are embedded in it (**e**). (**f**) At the end of the intradural dissection, a free CSF passage is established across the arachnopathy. Note the thin arachnoid layer left on the spinal cord covering its vessels. (**g**) A duraplasty has been inserted and lifted up with tenting sutures to enlarge the subarachnoid space limiting reobstruction by postoperative scar formation

ligaments to avoid injuries to motor pathways and anterior spinal cord vessels. Closing the microsurgical part of the operation, an expansile duraplasty is inserted with a tight running suture and finally lifted up with tenting sutures on either side (Fig. 9.8f). To avoid scar formation and tethering between duraplasty and the spinal cord, alloplastic material for duraplasty should be preferred, i.e., Gore-Tex® (W.L. Gore & Associates GmbH, 85640 Putzbrunn, Germany). Special attention is finally paid to a good, tight closure of the muscle layer to prevent any CSF from entering the epifascial space. In patients who have been operated before, as in patients with posttraumatic syringomyelia who underwent spinal instrumentation, for instance, a lumbar drain is placed prophylactically if the soft tissue appears scarred and sparsely vascularized.

Considerable experience is needed to be successful with this surgical technique. The more focused the surgery, the less scarring may result. If unnecessary steps are taken, such as a too extensive dura opening, or the surgical field is contaminated with considerable amounts of blood, postoperative scarring may counterbalance completely the effect of surgery. On the other hand, if the dura opening is not extensive enough to gain access to the normal subarachnoid space above and below the level of scarring, the procedure is insufficient. As always, it is the right measure that counts and determines whether an operation will be successful or not.

For patients with more extensive arachnopathies after meningitis (Fig. 9.7), multiple intradural surgeries, or spinal subarachnoid hemorrhage, for example, surgery cannot provide a normal CSF passage anymore [22, 25]. Axial MRIs taken from the entire area of the arachnopathy should be evaluated in such instances for evidence of cord compression. Quite often, pouches and cysts have formed causing profound cord compression over a few spinal segments. Such compressions can be treated surgically by a wide fenestration of the corresponding arachnoid membranes. Such an operation can improve neurological symptoms related to the cord compression, but it will not influence the syrinx.

For that purpose, thecoperitoneal shunts have been introduced, which drain CSF from the subarachnoid space above the level of obstruction to the peritoneal cavity [29, 36, 45, 47, 48] (Fig. 9.7). For cavities extending into the cervical cord, ventriculoperitoneal shunts have been used for the same purpose [38, 50]. However, these shunts have their problems. There is little experience concerning the correct pressure settings other than to set them as low as possible avoiding signs of overdrainage or low intracranial pressure. Programmable shunts are sometimes used, but the shunt systems available are not specifically designed for this purpose. No data exist, as to how much tissue coverage may be allowed over the valve in order to still be able to change the setting with the programming device. This leaves the problem where to position the valve. Low-pressure valves have been used to overcome these problems. Five patients in the author's series were treated that way. In one patient, a low-pressure valve was still not low enough, so that the valve was removed leaving the patient with a valveless drain. This worked for a year after which the catheter got blocked. Two patients do well clinically with a low-pressure valve even though the syrinx did not regress. The remaining two patients demonstrate a profound decrease of the syrinx with a favorable clinical response (Fig. 9.7).

For patients with a complete cord lesion, cordectomy is a very effective form of treatment for syringomyelia [6, 9, 15, 30, 41, 49] (Fig. 9.9). All patients treated in

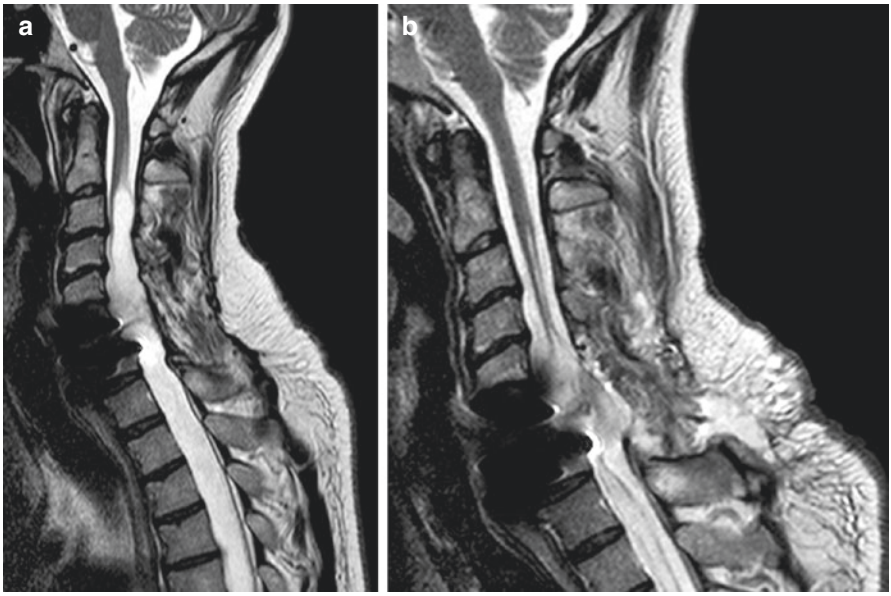

Fig. 9.9 (**a**) This T2-weighted MRI scan shows a huge posttraumatic syrinx almost 20 years after a complete cord lesion due to a dislocated C7 fracture in a 46-year-old man with progressive motor weakness of his hands. A first surgical attempt with arachnolysis and duraplasty at C6/7 stabilized the neurological status for 30 months. (**b**) The postoperative scan after cordectomy at C7 demonstrates the complete collapse of the syrinx

this manner in the author's series improved neurologically with permanent resolution of the syrinx. However, the psychological burden for a patient to accept this operation should not be underestimated. Most patients prefer to undergo a decompression first. After all, this operation does provide good results in the majority of patients [9]. Patients will accept a cordectomy, however, if the ascending neurology cannot be arrested by decompression or shunting procedures and the neurological progress threatens important functions such as respiratory or hand muscles.

9.4 Results

Concentrating on patients with syringomyelia related to spinal arachnopathies, 150 patients with posttraumatic arachnoid scarring and 333 patients with nontraumatic arachnopathies were encountered in the author's series. Reserving surgery for patients with progressive neurological symptoms led to operations for 73 patients with posttraumatic and 103 patients with nontraumatic arachnopathies (Table 9.2). Overall, 190 decompressions aiming at improving CSF flow and decompressing the spinal cord by resecting arachnoid pathologies were performed, while 11 patients with complete paraplegia underwent cordectomies, and 5 thecoperitoneal shunts were implanted. One patient received an opiate pump for

Table 9.2 Operations for spinal arachnopathies

Type of surgery	Posttraumatic arachnopathies	Nontraumatic arachnopathies	All
Decompression	69	121	190
Cordectomy	10	1	11
Thecoperitoneal shunt	1	4	5
Ventral fusion	5	4	9
Posterior fusion	7	4	11
Opiate pump	1	–	1

Table 9.3 Neurological recurrence rates for spinal arachnopathies

Patient group	5 years (%)	10 years (%)	*P*
All	34.2	52.4	
Nontraumatic focal	20.4	31.1	<0.0001
Nontraumatic extensive	67.3	72.8	
Nontraumatic all	35.2	44.9	
Posttraumatic no cord injury	22.0	22.0	n.s.
Posttraumatic incomplete cord injury	39.9	73.7	
Posttraumatic complete cord injury	32.0	54.6	
Posttraumatic all	31.8	60.9	

his neuropathic pain syndrome. The remaining operations dealt with degenerative diseases of the cervical spine.

Concentrating on the 190 decompressions with arachnolysis and duraplasty, complications were observed in 18.5 %. The most common were wound infections in 4.2 % and postoperative urinary tract infections in 4.8 %. CSF fistulas were observed in just 1.8 %. Permanent surgical morbidity defined as permanent neurological worsening within 1 month after surgery occurred after seven operations, i.e., 3.7 %. A postoperative decrease of the syrinx was observed in 68.5 %, 25.9 % showed no postoperative change, while 5.6 % increased further despite surgery. After 3 months, 51.5 % considered their condition improved, 40.1 % as unchanged, and 8.4 % as worsened. Looking at individual symptoms revealed postoperative improvements for sensory deficits and pain, whereas motor weakness, gait, and sphincter functions were left unchanged.

Long-term results were determined with Kaplan-Meier statistics to determine the rates for progression-free survival after decompression. Overall, 65.8 % remained in a stable neurological status for 5 years after surgery. This rate was reduced to 47.6 % after 10 years. Looking at particular subgroups revealed good long-term results for patients with a focal nontraumatic arachnoid pathology not exceeding two spinal segments (Fig. 9.6) and for posttraumatic patients who had conceded no spinal cord injury at the time of the accident (Table 9.3) (Fig. 9.7). For these subgroups, significantly lower clinical recurrence rates were determined after 10 years.

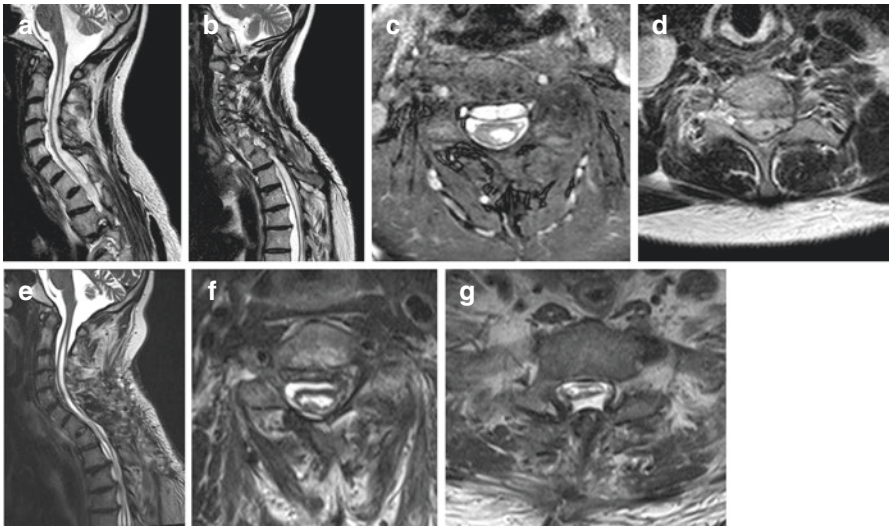

Fig. 9.10 (**a**) This T2-weighted MRI scan of a 53-year-old man shows a posttraumatic syrinx C2–Th5 related to a spinal injury at C7/Th1, which had resulted in an incomplete spinal cord injury 18 years earlier. The syrinx was caused by arachnoid scarring at the injury level combined with a narrowing of the subarachnoid space due to a posttraumatic dura dissection between inner and outer layer of the dura anteriorly from C2 down the entire thoracic spine (**b**). (**c**) This axial scan at C6 shows the syrinx in the posterior left of the cord. The dissection is caused by a rupture of the inner dural layer in the root sleeve C6/7 on the left side (**d**). At surgery, the CSF passage was established at the cervicothoracic junction, and the dura defect in the root sleeve was closed with muscle and fibrin glue. Postoperatively, the sagittal T2-weighted MRI demonstrates a reduction of the syrinx as well as the dura dissection (**e**). The axial scans at C6/7 (**f**) show fibrin glue in the left root sleeve and the dissection anteriorly as well as the duraplasty and free CSF space at Th1 (**g**)

Patients with extensive arachnoid pathologies or those with an incomplete spinal cord injury (Fig. 9.10), on the other hand, are the most difficult patients with spinal arachnopathies to treat. For patients with extensive arachnopathies after meningitis or intradural hemorrhages, the surgical concept of arachnolysis and duraplasty is as problematic as any other form of surgical treatment.

Thecoperitoneal shunts may reduce the ascending syrinx, but they hardly influence the myelopathy for which the arachnopathy itself is responsible (Fig. 9.7). The same applies to syrinx shunts. For patients with posttraumatic syringomyelia after an incomplete cord lesion, compromises as how far the arachnolysis and untethering of the cord should be pursued are unavoidable if surgical morbidity risking the remaining spinal cord functions is kept to a minimum (Fig. 9.10). It remains to be seen whether long-term results will improve with further experience. For patients with a posttraumatic syringomyelia who had suffered a complete spinal cord lesion at the time of their accident, results are more favorable, because a complete untethering and arachnolysis at the injury level can be performed without risking further neurological deficits (Fig. 9.11). If new symptoms appear postoperatively and the syrinx expands again, a cordectomy can always be performed later on (Fig. 9.9).

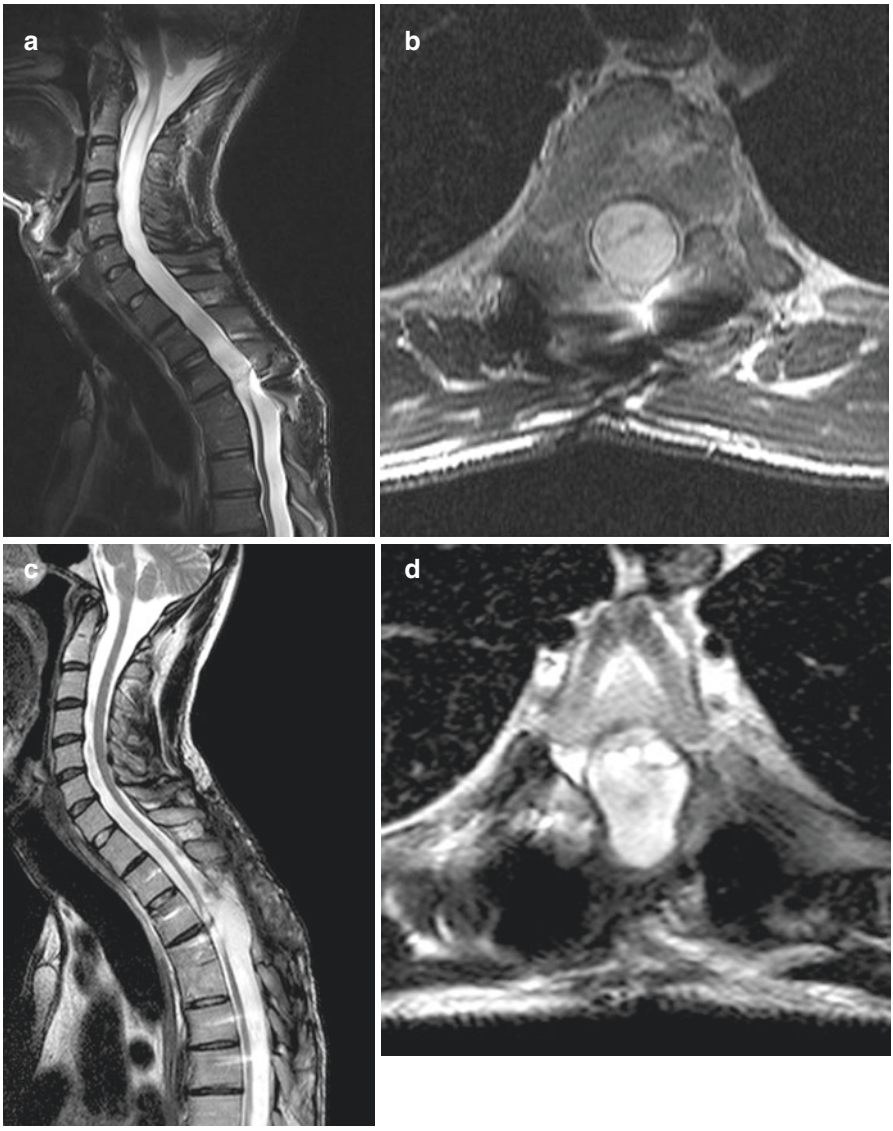

Fig. 9.11 (**a**) T2-weighted MRI scan of a 28-year-old man 6 years after an accident with fracture at Th4 and a complete spinal cord lesion. A huge syrinx has formed from C1 to Th4. (**b**) The axial scan at Th4 demonstrates the compression of the cord from posterior. After arachnolysis and duraplasty at Th4, the postoperative sagittal (**c**) and axial (**d**) scans show the complete collapse of the syrinx and the free CSF pathway at the injury level

Conclusion

The diagnosis of syringomyelia should be reserved for patients with a space-occupying intramedullary cyst of progressive character and differentiated from such entities as a dilatation of the central canal or myelomalacia [13, 25, 40]. Syringomyelia is not a disease in its own right but a manifestation of a disorder

of the spinal canal or craniocervical junction that has either resulted in an obstruction of CSF flow or spinal cord tethering or is associated with an intramedullary tumor. Management of patients with syringomyelia requires the correct diagnosis of the underlying disorder and the successful treatment of it. As this can be done in the vast majority of patients, no further surgical measures for the syrinx are required. Shunting the syrinx in particular can and should be avoided as the first line of treatment. The long-term prognosis depends on the therapeutic outcome of the underlying disorder.

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