Combined Surgical Treatment of Post-traumatic Syringomyelia: Case Report

A. Landi 1, P. Rocco 2, A. Ruggeri 3, N. Marotta, M. Cappelletti 4, R. Delfini 5

1,2,3,4,5 Department of Neurology and Psychiatry – Division of Neurosurgery, University of Rome Sapienza, Italy
dott. alessandro.landii@gmail.com

Abstract – Male 29 years, treated surgically for a traumatic fracture of L2, was referred to us about a year after for the onset of worsening paraparesis, anesthesia in his left arm and trunk, urgent incontinence, hyperhydrosis. An MRI of the spinal cord showed the presence of a holocord syrinx, extending to the filum terminale. The patient underwent surgery via L2 laminectomy, with lysis of the arachnoidal adhesions and resection of the filum terminale. After 14 months, MRI control highlighted the reduction of the syrinx and the improvement of symptoms. In post-traumatic syringomyelia the most effective surgical strategy is a combined treatment.

Key words- Post Traumatic Syringomyelia; Filum Terminale Section; Surgery; Combined Approach; Spine

I. INTRODUCTION

Post-traumatic syringomyelia is a major cause of delayed neurological deterioration, in patients with spinal trauma[1]. Two syndromes have been described as possible cause of major spinal injury: the Progressive Post-traumatic Cystic Myelopathy (PPCM) and the Progressive Post traumatic Myelomalacic Myelopathy (PPMM). The PPCM is a progressive pathological process that triggers the formation of a syrinx. The PPMM is due to microcystic degeneration of the spinal cord rather than formation of a syrinx[2,3]. Traditionally, syringomyelia has been treated by cystoperitoneal or cysto-subarachnoidal shunting, with limited effectiveness and with a 50% rate of clinical improvement[6,8]. Alternative treatments like lysis of subdural adhesions at the site of the trauma, opening of the syrinx and expansive duroplasty have been described. However, the results obtained with these techniques are similar to those obtained with shunts[9]. There is a need to improve the treatment of post-traumatic syringomyelia using a combined surgical approach which takes into account all the different pathogenic mechanisms. We present the case of a PPCM characterized by a holocord syrinx, treated by a combination of two different surgical techniques.

II. PATIENTS AND METHODS

A 29 year old male came to another Hospital, with a burst fracture of L2 following a motorcycle accident, with a severe motor deficit of the right lower limb was treated by L1-L3 screw fixation with posterolateral bone fusion, without decompressive laminectomy. The patient in the immediate postoperative course presents a persistence of the right lower limb motor deficit, and underwent physiokinesitherapy. The patient improved the neurological status until autonomous deambulation was achieved, but at 12 month follow-up the motor disorder has gradually worsened, interfering with deambulation. Other disorders like urgent incontinence, loss of sensitivity in the upper limbs, thermic and pain anesthesia, spasticity and numbness in the legs, sphincter dysfunction and hyperhydrosis also appeared. The patient underwent a lumbarosacral CT scan, which showed the correct positioning of the stabilization system. So he came to our attention and we performed an MRI of the whole spinal cord, that showed the presence of a large and deep syrinx with several compartments, extending from C2 to the conus medullaris, without any emerging points (Fig. 1). Based on the literature evidence and our experience in treating syringomyelia, we planning surgery combining two different surgical techniques: decompressive laminectomy (with the aim of spinal cord decompression) and filum terminale section (with the aim of mobilizing the medulla to remove cord tethering). So the patient underwent surgery treatment firstly by L2 decompressive laminectomy and secondly by neurolysis of the arachnoidal intradural adherences between the roots of the cauda, followed by a resection of the filum terminale and opening of the syrinx, (that included the filum terminale) (Fig. 2). The patient in the postoperative course, immediately improved his neurological status. Neuroradiological follow-up (performed immediately after surgery and at 1 and 6 months after surgery) showed a reduction of more than 50% of the volume of the syrinx (Fig. 3). MRI scan 14 months later showed a further reduction of the syrinx, whose volume had diminished by more than 90% of its pre-operative size (Fig. 4).

Fig. 1: Sagittal T2W MRI images showing the presence of a large and deep syrinx with several compartments, extending from C2 to the conus medullaris.
Fig. 2: Intraoperative image, that shows the opening of the cyst at the level of filum terminale

Fig. 3: Sagittal T2W MRI image showing the reduction of the volume of the syrinx at 1 month follow-up

Fig. 4: Sagittal T2W MRI image showing the further reduction of the volume of the syrinx at 14 months follow-up

A clinical evaluation at 24 month follow-up confirmed the progressive neurological improvement, with complete remission of hyperhidrosis, restoration of sphincter function, remission of numbness in the limbs and recovery of autonomous deambulation with a unilateral support.

III. DISCUSSION

Post-traumatic syringomyelia is an uncommon condition, but it is the most common form of syringomyelia after that due to malformative diseases. The incidence is 10% (of all spinal trauma), but recent MRI and autopic series have shown that this incidence may reach 20% an higher incidence between 22.5% to 53% is associated with a tethered cord mechanism. Trauma is the first cause of syringomyelia; It produces: hematomeylia, direct damage to the spinal cord and primary or secondary ischemic events, as inducing an inflammatory reaction, which promotes the formation of a syrinx. The products of bleeding and hemoglobin degradation induce inflammation, that leads to fibrosis within the spinal subarachnoid space. The adhesions causes an obstruction on the normal cerebrospinal fluid flow.
(from the ependymal canal toward the subarachnoid space), causing an inverse gradient of pressure. Consequently, increased resistance, produces a transmural CSF flow through the spinal cord. Variations in pressure that daily occur within the CSF system, are the primary mechanisms responsible for augmenting and maintaining syringomyelia, because they increase the latency necessary to achieve a physiological balance of pressure gradients between the CSF compartments. The syringomyelia grows in size after its formation. In our case the syrinx extension to the entire cord was probably attributable to late diagnosis and treatment. The traction exerted on the cauda equina and filum terminale by the subdural fibrosis, caused the extension of syringomyelia below the level of the fractura.

Post-traumatic syringomyelia is generally extracanalar, does not communicate with the central canal of the spinal cord and is not covered by ependyma, but by reactive gliosis. Furthermore, flexion-extension of the neck, causing traction on the spinal cord, could generate additional forces that complicate tense damage to the spinal cord. In these kind of patients the worsening of symptoms is due to spinal cord damage from pressure and tension determined by growth of the syrinx, as well as to local microvascular damage.

Hence et al, in cases of post-traumatic tethered cord, they suggest that traction exerted by the filum terminale in the spinal cord may play a role in the development and maintenance of syringomyelia. According to Williams, the deformations resulting from fractures may be also involved in the genesis of syringomyelia. Nowadays, neither the level of trauma nor other clinical and radiological evidence can predict which patients will develop syringomyelia and which will not. The symptoms caused by syringomyelia shall occur over a period of 3 months to 41 years after trauma with an average of 5-7 years. Clinical symptoms include root pain, neurological deficits, spasticity, and autonomic dysfunction.

Owing to the many underlying mechanisms, syringomyelia requires a complex therapeutic approach. Outcome is not always favorable, even when the results of radiological imaging are good. Currently, the main goal of treatment of syringomyelia is removal of the underlying cause: if this cannot be identified, shunting of the syrinx should be carried out. In the series described by Batzdorf, 36% of patients required more than one operation to solve syringomyelia. Historically, treatment of syringomyelia was based on a volume reduction of the syrinx in order to restore the flow of cerebrospinal fluid inside. The concept is based on the evidence that there is a correlation volume reduction of syringomyelia and better neurological outcomes. Surgical treatment consists of a cyst-subarachnoidal or cyst-peritoneal shunt, using the minimally invasive technique recently proposed by O’Toole. However the effectiveness of such treatment does not exceed 50%. Instead in all cases of posttraumatic syringomyelia the surgeon has to consider the cisto-peritoneal shunting option, but it’s low effectiveness means that it’s considered a second-choice. The cyst-subaracnoid or peritoneal shunting do not interrupt the pathogenetic mechanism of pathology, and for this reason it could be the cause of low effectiveness. The same authors underline the high incidence of shunt occlusion or dysfunction in a long term follow-up. Possible complications of shunting are arachnoiditis and infections, in addition to the presence of pre-existing spinal cord damage due to the trauma or tension exerted by syringomyelia. The clinical response to these treatments mainly concerns the painful symptoms and only marginally motor disturbances. Another therapeutic approach is decompressive laminectomy and/or revision of fixation systems, especially in cases where syringomyelia is caused by deformation or post-traumatic stenosis of the spinal canal, with consequent mechanical traction on the spinal cord.

In cases of post-traumatic syringomyelia secondary to a CSF dynamics variations in the sub-arachnoidal and subdural spaces, lysis of subdural adhesions with opening of the cyst and expansive duroplasty, represent the best approach, to restore a proper CSF flow with reduction of syringomyelia. Furthermore resection of the filum terminale in post-traumatic tethered-cord has proved to be a useful technique in association with these treatments. These treatments have proven effective both individually and in combination with techniques such as shunting, and the results obtained were the same as those observed after shunting alone. Combined treatment can act on multiple pathogenic mechanisms: resection of arachnoidal adhesions, elimination of the barrier impeding flow of cerebrospinal fluid, at the same time the shunt diverts CSF from the syrinx reducing its volume. In our case, several mechanisms contributed to the maintenance and progression of syringomyelia. The presence of fibrosis at the level of the cauda caused a tethered cord mechanism and impeded the CSF flow from the subarachnoid space through external spaces. Nonsurgical decompression of the spinal canal during the first surgery resulted in a canal stenosis. This condition made it necessary to perform multiple combined surgical techniques: a decompressive laminectomy and a removal of any arachnoidal adhesions and resection of the filum terminale. During surgery we found that the dura mater provoked fibrous adherences only with filum terminale, but wasn’t involved in epidural adhesions or interferences with the roots of cauda equina and didn’t cause any dural stenosis as it usually do in trauma. The roots and filum terminale were involved themselves in arachnoidal adhesions, so dura mater didn’t cause any CSF flow interferences. Therefore, we did not perform expansive duroplasty, due to the increased risk of pseudomeningocele and dural fistula. Single resection of filum terminale and root debridement was considered sufficient. In particular, during the section maneuvers of the filum terminale simultaneous opening of the cyst has been observed, because it involved this portion of spinal cord too. In fact, we may assume that a single treatment would not have been sufficient for resolution of the syringomyelia as suggested by Batzdorf. The combination of more than one method is, in our opinion, more effective: such methods include shunt techniques, because they make it possible not only to divert the fluid contained in the syrinx, but also to interrupt the mechanisms involved in the training and enhancing the syringomyelia.
IV. CONCLUSIONS

The traditional approach to post-traumatic syringomyelia, not only causes a variety of complications, but it is also uneffective in terms of clinical results in the long term follow up. Resection of the arachnoidal adhesion particularly in association with resection of the filum terminale eventually followed by expansive duroplasty, represents a valid and effective treatment strategy for post-traumatic syringomyelia, to reduce pressure in the syrinx and to remove any dorsal fibrosis. The decrease in tension stops further clinical progression or bring about an improvement in patient's neurological status. Combined surgical management represents an appropriate approach for effective treatment of posttraumatic syringomyelia, although the results obtained so far need to be supported by long-term follow-up studies. Considering the high possibility of recurrence, the patient should be strictly monitored in the time.

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