Case Report

Syringobulbia: A delayed complication following spinal cord injury – case report

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Context: Syringobulbia is a very rare progressive disorder of central nervous system, with several possible underlying conditions. Rarely, it is also encountered as a late complication of syringomyelia.

Findings: In the present manuscript, a case of a paraplegic patient, due to traumatic spinal cord injury (thoracolumbar fracture), presenting after years progressively developing symptoms of the lower cranial nerves and upper extremities, owed to syringomyelia and syringobulbia, the surgical treatment applied and its outcomes are described. We performed a syngo-peritoneal shunting procedure using a T-tube. The patient's symptoms resolved postoperatively and the cavity's size was reduced to a great degree.

Conclusion/Clinical Relevance: The late appearance of cranial nerve deficits or symptoms—signs of the upper extremities in a patient with traumatic thoracic spinal cord injury should raise suspicion that post-traumatic syringomyelia or syringobulbia has occurred. In such cases, radiologic evaluation and early surgical drainage of the cyst as a means of preventing significant delayed neurologic deficit is advocated.

Keywords: Syringobulbia, Syringomyelia, Syngo-peritoneal, T-tube, Cerebrospinal fluid shunt

Introduction

Syringobulbia is a very rare progressive disorder of central nervous system which is characterized by the formation of an elongated fluid-filled cavity, called syrinx, in the brainstem.^{1,2}

According to mythology, Syrinx was a nymph, who pursued by the Greek god Pan, ran to a river's edge and asked for assistance from the river nymphs. In answer, she was transformed into hollow water reeds which made a haunting sound when the god's frustrated breath blew across them. Pan cut the reeds to fashion the first set of "Pan pipes", which were thenceforth known as syrinx.³

While syringobulbia may occur in isolation, it may also occur secondary to neoplasms, spinal cord (SC) traumas or lesions, deformity of the craniocervical junction (Chiari malformation), meningitis, and other etiologies.^{4,5} Syringobulbia can be also rarely encountered as a late complication of syringomyelia, as the cavity extends toward the brainstem.^{4,5}

In this article a case of a paraplegic patient with syringobulbia developed years after a SC injury, due to the

expansion of a post-traumatic syringomyelia (PTS), the surgical treatment applied and its outcomes are analysed. This case report conforms to the CARE guidelines for case reports (<http://www.care-statement.org>).

Case report

History and clinical examination

A 40-year old male presented to our hospital with progressively developing symptoms over the last 4 years, which consisted of 1) sensory loss, concerning pain and temperature sensation, affecting the right upper extremity, 2) muscle wasting and weakness, concerning especially the most delicate movements of the right hand (MRC: Hand: 4-/5, Rest upper extremity: 4/5), 3) dysphagia (worse with fluids) and a subjective feeling of drowning, 4) dysphonia with the form of stuttering and hoarseness, 5) palsy of the right hypoglossal nerve, 6) hypoesthesia of the right side of the face, 7) hyperhidrosis of the right half of his body, as well as, 8) dyspnea. He mentioned that the sensory symptoms of the upper extremity worsened with violent coughing.

Fourteen years before his admission, the patient became paraplegic, due to a traumatic SC injury of the thoracolumbar junction. At that time, he underwent a surgical procedure of spinal instrumentation – fusion

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of the T11-L1 vertebral levels. Images from that admission are not available. On his current admission, magnetic resonance imaging (MRI) of the spine demonstrated a huge cyst extending from the T11 vertebral level to the medulla oblongata (Fig. 1). The cyst appeared as a well-circumscribed intramedullary fluid-filled mass. Therefore, the diagnosis of syringobulbia due to the upward extension of traumatic syringomyelia was obvious.

Operation

We performed a syringo-peritoneal shunting procedure using a T-tube. The patient was placed in a prone position. We chose the T1 vertebral level for the myelotomy site, because it was the widest portion of syrinx and the thinnest portion of the SC; thus, a site with the least potential to produce neurological deficits. After the T1 laminectomy and incision of the dura mater and the arachnoid membrane, we made a small midline myelotomy and opened the cyst. Then, a T-tube was placed at this level which was connected to a shunt tube cut to the desired length (one arm was inserted into the syrinx with rostral direction and the other arm with caudal direction). After a tight closure of the dura, we connected the T-tube with a catheter passed subcutaneously into the peritoneal cavity using the standard technique.

Follow-up

On his neurological examination at 6, 12 and 24 months after the operation, muscle strength (MRC: 5/5) and sensory function of the right upper extremity were

normalized. In addition, swallowing and facial hypoesthesia were considerably improved, too. Three magnetic resonance image (MRI) scans performed post-operatively showed reduction of the cavity's size to a great degree (Fig. 2). A timeline of the most important information of the patient's history, evaluation, treatment and outcomes is presented in Table 1.

Discussion

Syringobulbia clefts due to the increased pressure of the cerebrospinal fluid (CSF) in the fourth ventricle must be differentiated from the ascending syringobulbia which can occur from upward impulsive fluid movements in a syringomyelia. It has been suggested that the bulbar features are due to the effects of pressure differences acting upon the hindbrain with consequent distortion of the cerebellum and brainstem, compression and/or interruption of sensory and motor nerve pathways, traction on cranial nerves or indentation of the brainstem by vascular loops.⁶

Several mechanisms trying to explain the formation of the syringobulbia with a concurrent syringomyelia have been proposed. One of them is the rupture of the cervical syrinx rostrally resulting in syrinx formation in the brainstem. Another mechanism is the dilation and propagation of the spinal central canal of the medulla oblongata and brainstem leading to syringobulbia.⁴

Above, we presented a case of syringobulbia, which was developed as a result of PTS. PTS is a relatively uncommon, but potentially catastrophic complication of SC trauma, with an incidence varying from 0.3 to

Figure 1 Preoperative MRI scan of the cervical spine demonstrating severe syringomyelia and syringobulbia (huge cyst extending from the T11 vertebral level to the medulla oblongata).

Figure 2 Postoperative MRI scan (at 6 months) of the cervical spine demonstrating impressive reduction of the cavity's size, which was accompanied with a great neurologic improvement.

3.2%.⁵ The interval between the spinal trauma and the development of syringomyelia varies remarkably, too, with reported time periods ranging from 2 months up to 32 years.⁵ The postulated mechanisms include the “slosh” theory referring to pulsatile pressure surges from the accumulated intramedullary fluid following injury, the “suck” theory consisting of upward peaks of negative pressure working via a ball-valve effect, and finally the theory supporting the coalescence of microcysts.⁷

The clinical presentation of syringobulbia depends on the site of the cavity and the affected structures. As the lesion involves the brainstem, the patient presents cranial nerve palsies and corticospinal signs, with an

often insidious onset.⁴ According to the related literature, the commonest symptoms are occipital headache, vertigo, dysphonia, dysarthria, trigeminal paresthesia or hypoesthesia to pain and temperature, diplopia, tinnitus, palatal and/or hypoglossal palsy.⁸ Other possible symptoms include: singultus (hiccups), nystagmus, oscillopsia, Horner's syndrome or dyshidrosis, hearing loss, scapular winging, and facial myokymia.^{4,8–14}

In the case of PTS, the clinical feature with the highest frequency of appearance in the literature is pain, which can possibly be triggered by straining, coughing, sneezing or other mechanical influences. The second commonest symptoms include sensory deficits, especially dissociation between pain and proprioception or concerning vibration and touch sensation. Motor deficits follow in frequency representing the third most common symptom of PTS.⁵

Regarding the neuroanatomical background of the aforementioned patient's clinical presentation the following plausible explanations can be provided. The concurrent existence of sensory loss, concerning pain and temperature sensation, of the right upper extremity and hypoesthesia of the right side of the face can be only explained by dysfunction of the left lateral spinothalamic tract (2nd order neuraxons) and the left trigeminal lemniscus. The muscle wasting and weakness, concerning especially the most delicate movements of the right hand, should be attributed to injury of the left corticospinal tract. The dysphagia and the dysphonia should be associated with dysfunction of the vagus (pneumogastric) nerve.

It is remarkable that syringomyelia can be responsible for dyshidrosis (hyperhidrosis and hypohidrosis), as well as other disorders of the autonomic nervous system. According to the literature, hyperhidrosis is caused by stimulation, and therefore, hyperactivity of sympathetic preganglionic neurons (SPGNs). At present, two possible mechanisms leading to the hyperactivity of the SPGNs have been suggested. The first mechanism claims that the SPGNs are directly stimulated by minimal tissue damage, while the second supports the idea that the damage caused to the inhibitory local interneurons located in the vicinity of the SPGNs, and their consequent disinhibition, is the responsible factor. In either case, the hyperactivity of the SPGNs exists as long as the disability is mild. However, as the disease progresses the hyperactivity gradually decreases, shifts to normal activity, and is finally substituted with hypoactivity due to the progressive and irreversible damage to the SPGNs.¹³

Furthermore, a difficulty in breathing is highlighted in this patient. There is data indicating that the

Table 1 Timeline of the most important information of the patient's history, evaluation, treatment and outcomes.

Timeline
1) Traumatic spinal cord injury of the thoracolumbar junction.
2) Spinal instrumentation – fusion of the T11-L1 vertebral levels.
3) 14 years later presented with progressively developing symptoms (over the last 4 years), i.e. of the right upper extremity: sensory loss, concerning pain and temperature sensation, muscle wasting and weakness, regarding especially the most delicate movements of the right hand (MRC: Hand: 4-/5, Rest upper extremity: 4/5), hypoesthesia of the right side of the face, concerning cranial nerves: vagus nerve (dysphagia, dysphonia and dyspnea), right hypoglossal nerve, and hyperhidrosis.
4) MRI scan of the cervical and thoracic spine demonstrated severe syringomyelia and syringobulbia, extending from the T11 vertebral level to the medulla oblongata.
5) Syngo-peritoneal shunting procedure using a T-tube
6) On his follow-up at 6, 12 and 24 months postoperatively, impressive reduction of the cavity's size (almost eliminated) while the patient's symptoms resolved. At 6, 12 and 24 months postoperatively, muscle strength (MRC: 5/5) and sensory function of the right upper extremity were normalized and swallowing and facial hypoesthesia were considerably improved.

combination of dysphagia and dysphonia in longstanding syringomyelia and syringobulbia predict likelihood of respiratory disturbances, especially during sleep. In contrast, respiratory problems neither seem to be caused by muscle weakness nor are correlated with the size of the cavity.¹⁵

It is now clear that the previously described entity can certainly have serious consequences for every paraplegic or tetraplegic patient as even small neurological deteriorations can result in significant impairment in terms of daily activities. Therefore, immediate surgical treatment should be performed in patients with neurologic impairment. Nevertheless, there are still different options regarding surgical methods.⁵

Depending on the cause and type of syringomyelia, the procedures designed to correct this condition vary including: posterior fossa decompression, spinal subarachnoid space reconstruction, myelotomy alone or accompanied with various CSF diversion procedures, i.e. lumbo-peritoneal, syringo-pleural, syringo-subarachnoidal, or syringo-peritoneal shunt; and bisecting the filum terminale.¹⁶ According to a recent systematic review concerning postinfectious and post-traumatic adult-onset syringomyelia, arachnolysis was the only surgical treatment associated with a longer duration until clinically symptomatic recurrence ($P = 0.02$).¹⁷ In addition, it is important that the problems related with the shunt systems are avoided with this procedure. On the other hand, a large prospective study concerning post-traumatic syringomyelia investigated the outcome of 61 operations for decompression with arachnolysis, untethering, and duraplasty at the level of the underlying trauma. The authors concluded that the treatment of patients with preserved motor functions (ASIA C and D) remains a major challenge and posed the question whether thecoperitoneal shunts would be a superior alternative for this subgroup.¹⁸

Regarding the shunt systems implanted in order to treat this condition, it should be highlighted that they are often unreliable and present limitations, which is the reason why shunting is not routinely performed. More specifically, high rate of shunt failure (with all types of shunt) resulting in recurrence of symptoms and poor long-term outcomes have been reported in the literature.¹⁹ The principal cause of these phenomena is shunt obstruction, usually due to proximal shunt obstruction caused by the ingrowth of glial tissue into the holes of the tube. Some efforts have been made towards the direction of solving this problem; for instance, T-tubes possessing many side holes on their arms. However, there are further restrictions regarding the use of shunt systems. Firstly, the rotational forces

of the T-tube can be detrimental to the SC. Secondly; shunt dislocation is another fundamental reason for shunt malfunction.¹⁶ Moreover, if the T-tube has to be removed, this is difficult to do without causing SC injury. Needless to say, that there are other possible complications, for example infection and SC compression.

This article illustrated the concept and technique of a particular shunting procedure: syringo-peritoneal shunt using a T-tube. Despite all its potential drawbacks, it also demonstrates many advantages. Firstly, CSF can freely communicate between two different spaces: the syrinx and the peritoneal cavity, according to the pressure gradient. Secondly, the migration of the shunt catheter can be prevented, because dural anchoring of the T-tube is easy and solid. All things considered, when a shunt is needed, it is believed that this procedure is the most advantageous and widely recommended method among the various shunting procedures.¹⁶

Conclusion

The late appearance of cranial nerve deficits or symptoms – signs of the upper extremities in a patient with preexisting neurologic deficits of the lower extremities (e.g. paraplegia) due to traumatic thoracic SC injury, should raise suspicion that post-traumatic syringomyelia or syringobulbia has occurred. Radiologic evaluation in these patients should include MRI scans of the cervical and thoracic spine and brain. Consideration of early surgical drainage of the cyst as a means of preventing significant delayed neurologic deficit is advocated.

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