Case Report

Syringomyelia as a presenting feature of shunt dysfunction: Implications for the pathogenesis of syringomyelia

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Abstract

The pathogenesis of syringomyelia continues to be an enigma. The patency of the central canal and its role in the pathogenesis of communicating syringomyelia continues to elicit controversy. The case reported here provides an opportunity to retest some of the hypotheses of syringomyelia. A 33 year old female presented with sensory disturbances over the left upper extremity and trunk and was diagnosed to have panventriculomegaly with communicating syringomyelia. She was initially treated with ventriculoperitoneal shunting. As there was no change in her neurological status following shunt, this was followed by foramen magnum decompression with excision of an arachnoid veil covering the fourth ventricular outlet. She had clinical and radiological improvement after foramen magnum decompression. Five months later she had reappearance of the symptoms of syringomyelia and was found to have shunt dysfunction and holocord syrinx. She improved following shunt revision. This case is being reported to highlight the following points: 1. In patients with communicating syringomyelia and hydrocephalus, shunt dysfunction can present with symptoms of syringomyelia without the classical clinical features of shunt dysfunction, 2. In patients with communicating syringomyelia, the central canal of the spinal cord acts as an “exhaust valve” for the ventricular system, and, 3. studies about the patency of the central canal are reviewed in the context of this case and the role of the central canal in the pathogenesis of communicating syringomyelia is reviewed.

Key words: Shunt dysfunction, chiari malformation, foramen magnum decompression, syringomyelia, central canal

INTRODUCTION

The pathogenesis of syringomyelia continues to elicit controversy despite decades of research. An ideal surgical treatment for syringomyelia continues to[1,2] elude the neurosurgical fraternity. This is mainly attributable to our deficiencies in the knowledge of the pathogenesis of syringomyelia. The case presented here provides an opportunity to retest some of the hypotheses of syringomyelia.

CASE REPORT

A 33 years old female noted abnormal sensations in her left upper limb of two months duration. She was evaluated by neurologists and was believed to have psychiatric illness. She was then referred to a psychiatrist and was treated for somatoform disorder for two years when she complained of worsening of sensory disturbances in the form of sensory loss in a “cape-like” distribution along with painless ulcers in the left hand. At this time, the imaging of the cervical spine was done and she was found to have chiari I malformation and
Syringomyelia. She was later referred to neurosurgery for further management. On admission to neurosurgery department, she had normal optic fundi, sensory impairment from C4 to T8 on the left side with small, trophic ulcers in the left hand. Her left hand grip was weak. MRI examination revealed panventricular hydrocephalus with Chiari I malformation along with syringomyelia extending from C1 to lower thoracic region. There was evidence of communication of fourth ventricle with the syrinx [Figure 1a and b]. The patient underwent a right ventriculoperitoneal shunt. Following shunt insertion, there was no change in the patient’s neurological status. CT scan showed decompression of the supratentorial ventricles with persistent dilatation of the fourth ventricle [Figure 1c] and hence, the patient underwent foramen magnum decompression and intradural exploration. An upper cervical laminectomy was also done for a possible syringopleural shunt, if necessary.

During intradural exploration, an arachnoid veil was found to be covering the foramen of Magendie. The arachnoid veil was opened which revealed a gush of CSF. Following this the cervical cord size was found to decrease intraoperatively and hence the planned syringopleural shunt was not done. Following foramen magnum decompression, the patient’s neurological status improved with significant improvement in sensations and healing of trophic ulcers. Two months after surgery, she went back to work as a school teacher. Follow up MRI revealed collapse of the syrinx and decompression of ventricular system. However, interestingly, a thin streak of CSF representing the central canal was found extending from upper cervical to lower thoracic regions [Figures 2 and 3]. This was found to be consistent with the MRI appearance of the central canal as described by Holly and Batzdorf\(^1\) and Petit-Lacour \textit{et al.}\(^2\). Five months later, she presented with a one week history of difficulty in walking and diminished sensations over the left upper limb and left side of the trunk. There was no history of headache, vomiting or transient obscurations of vision.

![Figure 1: (a) Preoperative T1 weighted sagittal MRI showing panventriculomegaly with communication of the fourth ventricle with the holocord syrinx, (b) Preoperative T2 weighted sagittal MRI showing panventriculomegaly with communicating syringomyelia, (c) Postoperative CT scan after ventriculoperitoneal shunting showing decompression of the lateral ventricles with disproportionate dilatation of the fourth ventricle.](image)

![Figure 2: Postoperative sagittal T1 weighted (left panel) and T2 weighted (right panel) MRI showing significant reduction in the size of the ventricles and syrinx; arrow points to the persistent central canal.](image)

![Figure 3: Postoperative sagittal T2 weighted MRI of the thoracolumbar region showing the persistent central canal (arrow).](image)

![Figure 4: (a) Sagittal T1 weighted MRI at the time of shunt dysfunction showing panventriculomegaly with holocord syrinx, (b) Sagittal T2 weighted MRI at the time of shunt dysfunction showing panventriculomegaly with holocord syrinx.](image)
vision suggestive of shunt dysfunction. Examination at this
time revealed normal optic fundi, diminished sensations from
C2 to T8 on the left side, Grade 4/5 power in lower limbs,
exaggerated knee and ankle jerks and bilateral upgoing plantars.
Her shunt was found to be nonfunctional. MRI at this time
revealed pan ventricular hydrocephalus with the fourth ventricle
communicating with the holocord syrinx [Figure 4a and b].
Patient underwent a revision of the ventriculoperitoneal shunt
with gradual resolution of symptoms.

DISCUSSION

The pathophysiology of syringomyelia continues to be an
enigma. No single surgical approach – foramen magnum
decompression, ventricular shunting, shunting of the syrinx
has produced uniformly successful results. Underlying our
limitations of clinical management of syringomyelia is an
incomplete knowledge of the mechanisms of the formation and
enlargement of syrinx. Oi and colleagues also showed
that isolated CSF compartments can develop after shunting
or holoneural canal dilatation. Oi and colleagues introduced the term “hydromyelic
hydrocephalus” on the basis of their experience with nine
cases of hydromyelia associated with hydrocephalus. They
noted four stages in the evolution of hydromyelia associated
with hydrocephalus: Stage I: dilatation of lateral ventricles,
Stage II: dilatation of lateral and third ventricles, Stage III:
dilatation of lateral and third ventricles with disproportionate
dilatation of fourth ventricle, and Stage IV: dilatation of all the
ventricles with communication of the fourth ventricle with the
dilated central canal. Oi and colleagues called stage IV as
Holoneural canal dilatation. Our case conforms to Oi’s Stage IV
or holoneural canal dilatation. Oi and colleagues also showed
that isolated CSF compartments can develop after shunting in
such patients. They showed that four different types of
isolated CSF compartments may occur after shunting due
to functional obstruction: Type I: when there is functional
obstruction of the foramen of Monro there is progressive
unilateral dilatation of one lateral ventricle; Type II: when
there is aqueduct stenosis along with obstruction of the fourth
ventricular outlets there is isolated fourth ventricle; Type III:
dilatation of the fourth ventricle which communicates with the
central canal of the spinal cord and this was termed as
“isolated rhombencephalic ventricle”and, Type IV: where
there is isolated dilatation of the central canal of the spinal
cord. Hall and colleagues in a series of elegant experiments have shown
that in communicating syringomyelia when the intraventricular pressure is increased there was increase in the intrasyringeal pressure. However, when the intraventricular pressure was decreased there was no corresponding decrease in intrasyringeal pressure which continues to remain elevated. They postulated that the absence of decrease in intrasyringeal pressure after reducing the intraventricular pressure was due to the existence of a “functional ventriculosyringeal valve” which opens in response to elevated intraventricular pressure and closes in response to decrease in intraventricular pressure. In their study, they found that a prolonged increase in intracranial pressure reduced the baseline ventricular pressure by increasing the intrasyringeal pressure and hence, they postulated that the syringeal pressure at rest may be maintained by displacement of ventricular fluid into the syrinx cavity during transient increases in ICP. They postulated that the ventriculosyringeal valve might be located at the level of the proximal central canal [Figure 5]. The existence of the ventriculosyringeal valve might explain the type IV isolated compartment (isolated dilatation of the central canal of the spinal cord) of Oi and colleagues.

Pathogenesis of syringomyelia in the index case

The pathogenesis of syringomyelia in our case can be summarized as follows: Stage I: the disease process started with panventricular dilatation [Figure 6a]; Stage II: persistence of this panventricular dilatation led to the development of communication between the dilated fourth ventricle and the central canal (holoneural canal dilatation) [Figure 6b]; Stage III: decompression of the supratentorial ventricles by shunting the lateral ventricle led

![Figure 5: (a) Schematic representation of panventriculomegaly without dilatation of the central canal of the spinal cord, (b) Schematic representation of the dilated ventricular system communicating with the dilated central canal of the spinal cord causing “holoneural canal dilatation” (Adapted from Oi et al.; J Neurosurg 74: 371-379, 1991)]]
to the formation of isolated rhombencephalic ventricle as suggested by Oi and colleagues as manifested by the fact the patient’s symptoms of syringomyelia did not improve even after shunting the lateral ventricles [Figure 7]; Stage IV: formen magnum decompression with excision of the arachnoid veil led to the decompression of the isolated rhombencephalic ventricle and the patient’s symptoms improved; Stage V: when the patient presented with shunt dysfunction, she reverted back to stage II with panventricular dilatation and holocord syringomyelia (i.e., holoneural canal dilatation).

Rapid clinical and radiological improvement following treatment of communicating syringomyelia with ventricular shunting has been documented by several reports as discussed below:

Ogilvy and Borges reported the rapid improvement in the symptomatology of communicating syringomyelia with associated hydrocephalus in a 58 year old man with a long history of symptoms. In a previous report, Milhorat and colleagues have shown that a patient with hydrocephalus, chiari I malformation and syringomyelia who had been treated with ventriculoperitoneal shunt and foramen magnum decompression one of the presentations of shunt dysfunction is rapid occurrence of syringomyelia. Milhorat and colleagues equated the dilated central canal in this condition to a “fifth ventricle”.

Peraud and Grau recently reported a 33 year old female who presented with postmeningitic hydrocephalus with communicating syringomyelia which rapidly improved following surgical intervention. It has been previously shown that when a patient with arrested hydrocephalus decompensates, development of a presyrinx state might be one of the ways of decompensation and treatment of the decompensated arrested hydrocephalus can reverse the presyrinx state. More recently, Camacho and colleagues have documented the development of communicating syringomyelia in a preterm infant with post hemorrhagic hydrocephalus. More recently, Hagihara and Sakata reported a 13 year old boy with disproportionately large communicating fourth ventricle with syringomyelia in whom there was complete clinical and radiological improvement following placement of ventriculoperitoneal shunt. The case reports by Peraud et al., Muthukumar et al., Camacho et al., and Hagihara et al. have shown even in the absence of chiari malformation communicating syringomyelia can develop rapidly and this syrinx responds well to CSF diversion. These case reports lend credence to the existence of the “potential” for communication between the fourth ventricle and the “potentially patent” central canal of the spinal cord (to be discussed below). If this is so, why then communicating syringomyelia does not develop in the commonly encountered condition of obstructive hydrocephalus in infants? This is because of the following reasons: 1. Many cases of obstructive hydrocephalus are due to aqueduct stenosis and hence the fourth ventricle is not dilated and, therefore, communicating syringomyelia does not develop 2. In those infants with obstructive hydrocephalus due to fourth ventricular outlet obstruction, the open fontanelles and unfused sutures act as an “exhaust valve” with resultant increase in head circumference and thereby prevent the development of communicating syringomyelia. The reasons for the infrequent development of communicating syringomyelia in adults will be discussed below.

This case highlights the following points:

- The pathophysiology of syringomyelia is multifactorial. Explanations for the development and maintenance of syringomyelia might vary from case to case and it might not be possible or even necessary to have a single hypothesis to explain the pathogenesis of all forms of syringomyelia.
- In cases where there is radiological evidence of patent communication between the syrinx and the “dilated” fourth ventricle, the development and maintenance of syringomyelia is by the flow of CSF from the ventricles into the central canal.
- Absence of improvement following ventricular shunting procedures does not militate against the existence of this “communicating” syringomyelia as this can be explained the ventriculosyringeal valve effect of Hall and the isolated rhombencephalic ventricle of Oi.
Patients with hydrocephalus and communicating syringomyelia when they deteriorate after treatment, shunt dysfunction might be heralded by the reappearance of syringomyelic symptoms without the presence of signs and symptoms of increased intracranial pressure as the development of syringomyelia in this condition is due to decompression of the ventricles into the central canal of the spinal cord. In such cases, the central canal of the spinal cord acts as an “exhaust valve” for the dilated ventricular system. Clinicians should be aware that shunt dysfunction might present with symptoms of syringomyelia in such patients without overt clinical signs of increased intracranial pressure.

Postoperative scans after foramen magnum decompression in our patient [Figure 2] showed the collapsed but persistent central canal. This highlights the fact that at least in some patients with communicating syringomyelia, once syringomyelia develops, the normal age-related obliteration of the central canal as suggested by Cornill and Mossinger, Kasantikul et al., Netsky et al., Milhorat and colleagues, Yasui and colleagues[21-24] does not happen and hence these patients are at a greater risk of syrinx recurrence. This is further reiterated by the case report of Milhorat et al.[25] where the syringomyelia reappeared eight years later during an episode of shunt dysfunction thereby showing that in such patients true age-related obliteration of the spinal canal does not happen even several years after treatment.

Misconceptions about the Stenosis of the Central Canal

• AUTOPSY STUDIES
The autopsy study by Milhorat and colleagues is often widely quoted to substantiate the age dependent stenosis of the central canal.[26] However, a detailed analysis of Milhorat’s study reveals many interesting facts. Milhorat et al. classified stenosis of the central canal as follows: Grade 0: No stenosis, Grade 1: 1-25% reduction, Grade 2: 25% to 50% reduction, Grade 3: 50% to 75% reduction, Grade 4: 75% to 99% reduction, and Grade 5: 100% reduction. In their study, Grade 5 occlusion of the entire canal was present only in four of the 232 cases and these individuals were 43, 59, 60 and 73 years of age respectively. This means that in the majority of individuals, even though, the central canal becomes stenosed it does not become totally occluded and thereby remains functionally patent. In the very same study, five adults older than 20 years had completely patent canals and 18 others had patent canals except for partial stenosis (Grade 1 to 3) at one or two levels. They also mentioned that even in individuals who had stenosis there was relative sparing of the rostral and caudal ends of the canal.

An excellent autopsy study by Yasui et al.[25] showed that even though the central canal gets progressively obliterated with age, the portions of the central canal in the cervical cord remain patent up to the fourth to sixth decades of life as is evidenced by a detailed analysis of their Figure 5. The concept of central canal patency received support from an autopsy study by Newman and colleagues who studied the spinal cord of 60 individuals without neurological illness.[26] They found that central canals exhibited great morphological variability ranging from an ill-defined core of ependymal cells to a patent canal upto 1000 µ in diameter. They also showed that in the second decade the central canal can remain patent throughout the length later becoming progressively occluded after the fourth decade onwards. They also mentioned that occasionally, the central canal can remain patent in the cervical and lumbar regions even in older individuals.[26] This is in accordance with the autopsy study by Milhorat and colleagues who found that the central canal often remains patent in the rostral and caudal ends even in older individuals.[24] The findings of Milhorat et al., Newman et al. and Yasui et al.[24,26] have the following implications for the pathogenesis of syringomyelia: In patients in whom the central canal remains patent, panventricular dilatation causes the diversion of the pent up CSF into the central canal and in those in whom the canal had become obliterated, the same does not open in response to ventriculomegaly. The failure of the occluded canal to open has been explained by Yasui and colleagues: Because of proliferation of ependymal cells and astrocytes.[25]

Clinical studies
A recent clinical study by Holly and Batzdorf has shown that in normal individuals, the central canal of the spinal cord can be visualized by MRI.[30] In their study, in the majority of the individuals, the appearance of the central canal remained static during follow up. Suprisingly, in accordance with the autopsy study of Yasui et al., the highest incidence of persistent central canal was found in the cervical region.[31] The mean patient age in their study was 40 which once again correlates with the autopsy study of Yasui et al.[25] An MRI study by Petit‑Lacour and colleagues showed that with 1.5 Tesla MRI imaging it is possible to visualize the central canal in 1.5% of normal individuals of whom 83% were females with a mean age of 34 years.[4] With the increasing availability of 3.0 Tesla MRI, the incidence of this finding is likely to increase. These studies further lend credence to the hypothesis that the central canal can remain patent in a proportion of individuals, most often in the cervical region, and thus may participate in the pathogenesis of syringomyelia.

• Certain authors do not accept the existence of functional communication between the fourth ventricle and the syrinx.[27] The case presented here and the case reports and the studies alluded to in the earlier part of the discussion prove that in certain patients with communicating syringomyelia, there is a functional communication between the ventricles and the syrinx. This case report does not negate the other theories of origin of syringomyelia. On the other hand, it highlights that the pathogenesis of syringomyelia is much more complex than can be explained by the theories propounded so far and it is not possible or even necessary to have a one-size-fits-all theory to explain all forms of syringomyelia. Failure to understand the difference in the pathogenesis of different types of syringomyelia can lead to inappropriate treatment with suboptimal results. Trying to explain the pathogenesis of syringomyelia by a single theory can satisfy Occam’s razor.[28] While satisfying Occam’s razor is a worthwhile goal in scientific pursuit, improper use of Occam’s razor can lead to a perpetuation and corroboration of existing prejudice, and therefore,
Occam’s razor should not be used to get rid of unwelcome data or concepts.\(^{[26]}\)

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