Syrinx Extending from Conus Medullaris to Basal Ganglia: A Clinical, Radiological, and Pathological Correlation

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ABSTRACT: A 41-year-old woman with a history of birth injury to the brachial plexus suffered several delayed episodes of neurological deterioration. Magnetic resonance imaging studies revealed a syrinx extending from the conus medullaris into the brainstem and rostrally into both internal capsules. She died of an acute exacerbation of chronic respiratory failure. Autopsy demonstrated syringomyelia and syringobulbia with cavity extension bilaterally along the corticospinal tracts into the internal capsules. Islands of glial tissue in the subarachnoid space around the medulla caused obstruction of the subarachnoid space at the foramen magnum. These were probably the result of birth injury to the cerebellum. A detailed clinico-pathological correlation is provided to explain her neurological deficits. The pathogenesis of syrinx formation is discussed in terms of a late manifestation of birth trauma.


Patients with syrinx extending above the medulla are uncommon. There are autopsy descriptions of two patients with a syrinx extending to the midbrain and four patients with a syrinx extending into the cerebrum, usually along the internal capsule. Recently there have been radiological descriptions of patients with syrinxes extending from the spinal cord into the internal capsule with damage to the putamen, thalamus, or corona radiata. In this report we present the clinical, magnetic resonance imaging (MRI), and autopsy findings of a patient with a syrinx extending from the conus medullaris into both internal capsules.

CASE REPORT

The patient was a 41-year-old right handed female whose presenting complaint was right side weakness. At birth in 1946 she presented in a breech position and was delivered vaginally. Following delivery she cried but was unable to move at the left shoulder or elbow. At age 9 years an operation was done to allow limited movement about the left shoulder. At age 15 years she experienced gradual onset of balance disturbance, slurred speech, difficulty swallowing, impairment of gaze to the left side, and left lower motor neuron facial weakness. Except for impaired abduction of the left eye and mild facial weakness, these deficits resolved over a period of approximately 2 months. A provisional diagnosis of multiple sclerosis was made at that time. Subsequently, a surgical procedure was performed to improve abduction of the left eye. She had two uneventful pregnancies. At age 36 years after lifting a heavy object she experienced, over the period of 1 week, progressive weakness of the left arm and leg which never resolved completely. At age 41 years, 8 months prior to hospital admission, she fell on her back and experienced pain radiating from left neck and back into the left upper arm, progressive impairment of speech volume and clarity, left arm and leg weakness, and unsteadiness of gait. As a result she was unable to continue her job as a typist. She had no bowel or bladder dysfunction, hearing loss, vertigo, nausea, vomiting, or visual deficit. She was on no medications but had smoked five cigarettes per day for many years.

Neurological examination revealed normal cognitive function. Olfaction was normal. Uncorrected visual acuity was 20/100 but pupillary function, visual fields, and ocular fundi were normal. Neither eye moved beyond the midline to the left side and the left eye did not fully adduct. Facial and corneal sensation and muscles of mastication were normal. There was lower motor neuron weakness to the left face. Hearing was normal. She had...
dysarthric speech, was unable to close her glottis, and had poor movement of the left palate. Shoulder shrug on the left side and both sternomastoids were weak. Her tongue deviated slightly to right side but there was no fasciculation or atrophy. All sensory modalities were impaired in the left hand and forearm and the left leg had decreased position and vibration sense. She had normal muscle bulk except for wasting of the intrinsic muscles and hypothenar eminence of the left hand. The right upper limb had normal strength. There was severe weakness of the proximal and distal muscles of the left upper limb with relative sparing of only wrist flexion. She had weakness of the left foot dorsiflexors and the left abdominal muscles. There was a slight increase in muscle tone on the left side. The left triceps and biceps stretch reflexes were diminished. Both legs were hyperreflexic with ankle clonus on the left. Both plantar responses were extensor. Her gait was slow but steady. She had left arm dymetria. The general physical examination was unremarkable except for a resting respiratory rate of 20 breaths per minute.

MRI examination (Figure 1) demonstrated a continuous irregular syrinx extending from the conus medullaris into the brainstem. A high signal intensity area on T2-weighted images was identified in the right basis pontis, right cerebral peduncle, and bilaterally in the basal ganglia. On the right side the area extended into the corona radiata. The cerebellum appeared to be impacted in the foramen magnum. There was partial fusion of the C4 and C5 vertebral bodies and significant thoraco-lumbar scoliosis. Plain radiograph of the spine showed a thoracic scoliosis convex to right with a lumbar compensation curve.

To further define the syrinx, contrast agent was injected into the spinal cord at the C1-C2 level. Computed tomography scan showed contrast in the subarachnoid space and syrinx but the extent of the cavity was not clearly visualized. Three days later she complained of increased weakness on the left side. Six days after the injection she was taken to the operating room for syringosubarachnoid shunting. Following administration of the anesthetic she could not be intubated despite the use of a

Figure 1 — a) Parasagittal T1-weighted magnetic resonance image demonstrating cerebellar tissue extending into foramen magnum (arrow) and a small CSF intensity space in the pons (arrowhead). b) Sagittal T1-weighted MR image demonstrating a large irregular cavity in the caudal medulla and spinal cord. The bodies of the fourth and fifth cervical vertebrae are partially fused. c) Horizontal T2-weighted MR image at level of pons demonstrating a high signal area in the right basis pontis (arrow). d) Horizontal T2-weighted MR image at level of basal ganglia demonstrating bilateral regions of CSF intensity signal lying adjacent to the posterior limbs of the internal capsules (arrows). The larger cavity is on the right side. The ventricles are of normal size.
fiberoptic bronchoscope. During manual ventilation the arterial 
P\text{CO}_2\text{ rose to } 20 \text{ kPa (normal range 4.7-6.0)}. Spontaneous respi­
ration returned and 45 minutes later the arterial \text{P\text{CO}_2} was 11.6 

kPa.

The following day movement of the left chest and diaphragm 
was noted to be poor and vital capacity was 0.5 litres. Despite 
\text{P\text{CO}_2} in the 9-11 kPa range she was alert and oriented and felt 
well. A diagnosis of chronic central hypoventilation with weak 
inspiratory musculature and a mild degree of lower airway 
obstruction secondary to cigarette smoking was made. Her res­
piratory drive appeared to be based on hypoxia alone. The patient 
demanded that she not be intubated for chronic mechanical 
ventilation. Using respiratory stimulants and controlled oxygen 
therapy her oxygenation slowly improved. Despite mild right 
side and severe left side weakness she was able to walk with 
assistance on the third post-anesthetic day. Two days later, how­
ever, she complained of increased weakness and difficulty 
breathing and the following day she was cyanotic. She died of 
respiratory failure. An autopsy was performed.

**Autopsy Findings**

The general autopsy revealed bilateral patchy bronchopneu­
monia. The skull was unremarkable. Because the MRI lesions in 
the basal ganglia were not originally interpreted as syrinx exten­sions, the brain was removed by transection through the mid­
brain. The dura mater, venous sinuses, leptomeninges, basal 
arteries, and external surface of the cerebral hemispheres were 
unremarkable except for asymmetry of the mammillary bodies 
and fibrous adhesions around the medulla. In spite of the scoliosis 
and fusion of the cervical vertebrae, the spinal canal had no evi­
dence of narrowing. The spinal dura and arachnoid were thick­
ened and adherent to the cervical and upper thoracic spinal cord. 
The rostral 6 cm of the cervical spinal cord were expanded. The spinal nerve roots of the lower cord were unremarkable. The syrinx was injected with barium contrast agent for postmortem 
radiography. The brachial plexus was not examined.

Coronal sections through the cerebral hemispheres revealed 
bilateral cavities located between the internal capsule and lenticular 
nucleus (Figures 2 and 3). The right side cavity was up to 0.3 cm 
wide and had a rubbery wall with yellow discoloration. It extended rostrally into the corona radiata and caudally to merge 
with a pinpoint cavity in the right cerebral peduncle. The left 
cavity was very narrow, had no discoloration of the wall, and 
could not be traced into the cerebral peduncle. The lateral and 
third ventricles were of normal size and there were no other 
abnormalities of the cortex, white matter, or basal ganglia.

Horizontal sections through the brainstem revealed a pin­
point cavity in the midbrain lying between the right cerebral 
peduncle and substantia nigra. This continued into the basis pontis, 
where it widened to 0.2 cm diameter, and then caudally into the 
medulla where the defect was located between the inferior olivary 
nucleus and the right pyramid. Multiple cavities were apparent in 
the lower medulla. The aqueduct and fourth ventricle were of normal 
size. Cross sections through the spinal cord showed an irregular, 
widely patent syrinx extending the entire length of the spinal cord.

**Microscopic Findings**

Tissue sections were stained with Luxol fast blue/hema­
toxylin and eosin. Selected blocks were sectioned serially to 
define the complex branching of cavities in the medulla. 
Selected sections were stained with Masson’s trichrome, PTAH, or 
for glial fibrillary acidic protein using the avidin-biotin complex 
method.

In the sacral spinal cord the syrinx was lined almost entirely 
by ependyma. In the lumbar and thoracic spinal cord the wall 
was composed of a dense layer of astroglial fibers which con­
tained buried clusters of ependymal cells and was covered only 
locally by islands of ependymal cells. The white matter columns 
were compressed and exhibited a slight decrease in myelin 
staining. The anterior white commissure was preserved. Despite 
distortion of the anterior horns, the motor neurons were spared. 
The spinal roots and dorsal root ganglia appeared normal.

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**Figure 2** — Photograph of a coronal slice through the brain showing 
the narrow cavity in the left internal capsule (ic) adjacent to the 
posterior putamen (p). g = lateral geniculate body.

**Figure 3** — Photograph of a coronal slice through the brain showing 
the cavity in the right internal capsule and corona radiata (cr). 
Narrow bridges of glial tissue span the lumen. The putamen (p) and 
globus pallidus are damaged focally. t = thalamus.
The entire cervical spinal cord was severely distended. All white matter tracts, especially the corticospinal tracts, showed gliosis and diminished myelin staining. The syrinx was up to 1 cm in diameter and at some points the gliotic wall was 1.5 mm thick and extended to the external surface of the cord. Focal islands of ependyma lined the cavity. The lower cervical levels exhibited asymmetric loss of anterior horn motor neurons with atrophy of the anterior roots. In the upper cervical cord the ependyma-lined central canal lay entirely between the syrinx and the anterior commissure (Figure 4). Although patent, the central canal was small and the exact level of convergence with the syrinx was not defined. A macrophage-lined needle tract corresponding to the CT-guided injection site was found at the C1-2 level. Fibrous thickening of the arachnoid surrounded the upper cord. Near the cervicomedullary junction were vascularized islands of glial tissue lying in the subarachnoid space.

At the cervicomedullary junction the syrinx divided into up to eight channels separated by glial septations (Figure 5). All were lined by a dense gliotic layer with scattered Rosenthal fibers but no ependymal cells. Only a few cavities contained barium contrast material. Some channels were separated from the subarachnoid space by only a delicate cell layer. The largest channels were located in the left anterior cord and medulla adjacent to the largely demyelinated decussation of the pyramids. Distal to the decussation, the right corticospinal tract was more richly myelinated than the left. There was moderate sparing of myelinated axons in the fasciculus gracilis and cuneatus and spino cerebellar tracts. The central canal was small and in its expected location. The caudal solitary nuclear complex, which is located posterior to the central canal and which receives major afferent connections from chemoreceptors and baroreceptors, was gliotic with a paucity of neurons. The area reticularis superficialis ventrolateralis, a major source of efferents to respiratory motoneurons in the spinal cord, appeared normal. The cortex of the left cerebellum at the level of the cervicomedullary junction was focally replaced by a dense astroglial scar. In the subarachnoid space adjacent to the scar were abundant islands of glial tissue (Figure 5). The lateral foramina of the fourth ventricle were patent.

Figure 4 — Photomicrograph of the upper cervical spinal cord showing the dense glial lining (between arrowheads) of the syrinx (S). The ependyma-lined central canal (curved arrow) lies between the syrinx and the anterior white matter commissure. Luxol fast blue hematoxylin and eosin. Bar = 0.5 mm.

Figure 5 — Whole mount section through inferior medulla showing numerous cavities primarily in the anterior left quadrant. The subarachnoid space on the left side is largely obliterated by ectopic islands of glial tissue (arrowheads) and the adjacent dura is thickened. d = decussation of the corticospinal tracts, v = vertebral arteries. Masson's trichrome. Bar = 2.5 mm.

The cavities in the medulla tapered rostrally. At the caudal limit of the pyramids there were only three irregular cavities all of which contained barium (Figure 6). One cavity was located in the left medullary tegmentum anterior to the inferior olivary nucleus and lateral to the hypoglossal nucleus and the well myelinated left medial lemniscus. The cavity and surrounding glial scar interrupted fibers from the left nucleus gracilis and cuneatus. The right medial lemniscus was devoid of myelinated axons. A broad area of gliosis obliterated the left nucleus ambiguus. The cavity ascended through the tegmentum and approached the floor of the fourth ventricle where it obscured the left motor nucleus of the vagus and the nucleus solitarius. In the tegmentum of the lower pons the cavity and surrounding gliosis obscured the left facial nucleus, left abducens nucleus, and medial longitudinal fasciculus. Just rostral to the facial nucleus the cavity communicated with the fourth ventricle through a narrow channel. Further rostrally the cavity collapsed to a narrow glial scar which faded in the mid pontine tegmentum. The fourth ventricle and aqueduct were unremarkable. The second medullary cavity was located between the right pyramid and inferior olivary nucleus where it interrupted axons of the right hypoglossal nerve. This cavity followed the corticospinal tract and enlarged as it ascended through the medulla into the basis pontis (Figure 7). In the cerebral peduncle it became narrow and difficult to follow. From the cerebral peduncle the cavity passed medial to the optic tract into the internal capsule where it was surrounded by a dense band of gliotic tissue and scattered hemosiderin-containing cells. There were foci of tissue destruction and gliosis in the adjacent medial globus pallidus and superior putamen. The third and smallest medullary cavity was located between the left pyramid and inferior olivary nucleus. In the mid-medulla it collapsed to a narrow glial band which ascended alongside a bundle of corticospinal tract axons through the pons into the midbrain, essentially mirroring the path of the cavity on the right side. In the left internal capsule the cavity was patent and surrounded by a narrow band of gliosis which seemed to be in continuity with the glial scar in the cerebral peduncle.
Clinico-pathological Correlation

The profound left arm weakness and sensory deficit were a direct consequence of this woman's birth injury. Unfortunately we were unable to obtain further details concerning her disability after birth. The injury was likely unilateral damage to the proximal upper brachial plexus with subsequent atrophy of the anterior nerve roots.13 Beginning at age 15 years she developed impairment of gait and swallowing, slurred speech, left facial weakness, and impaired leftward gaze. In retrospect these deficits were probably caused by syringobulbia which went unrecognized. They can be explained by rostral extension of the syrinx from the medulla into the left pontine tegmentum with involvement of somatosensory fibers from the left body, the motor nucleus of the vagus nerve, the nucleus solitarius, the facial nucleus, and the abducens nucleus. The gait impairment may have been due to interruption of the spinocerebellar tracts in the medulla or cervical cord or the corticospinal tracts at their decussation. The resolution of signs may have followed rupture of syrinx into the fourth ventricle. As is demonstrated in this patient, the presentation of syringobulbia may be insidious and can be mistaken for other disorders such as multiple sclerosis.1,14

At age 36 years, following strenuous exertion the patient experienced further weakening of the left arm and leg followed by only partial recovery. These findings could be accounted for by extension of a syrinx along the right corticospinal tract into the internal capsule with focal hemorrhagic tissue destruction. Finally, subsequent to a fall 8 months prior to death she experienced further impairment of gait and speech. The neurological examination findings can be explained by the location and extent of the syrinxes. The bilateral leg hyperreflexia reflects injury to the corticospinal tracts at their decussation or along the length of the spinal cord. The loss of position and vibration sense of the left leg reflects the near complete loss of fibers projecting from the left nucleus gracilis and cuneatus to the right medial lemniscus. Interruption of the right hypoglossal nerve axons near their site of exit between the pyramid and the inferior olive caused deviation of the tongue to the right and dysarthria. Weakness of the sternomastoids and respiratory musculature was likely due to severe distention of the upper cervical spinal cord. Damage to the caudal solitary nuclear complex with loss of ability to process input from chemoreceptors probably eliminated the hypercarbic respiratory drive. This has previously been reported in a patient with syringobulbia.15 The gaze abnormality is characteristic of a left side one-and-one-half syndrome which is a combination of ipsilateral horizontal gaze palsy and internuclear ophthalmoplegia. It was caused by the syrinx in the left pontine tegmentum which damaged the abducens nucleus, the medical longitudinal fasciculus, and the paramedian pontine reticular formation.16,17

Pathogenesis

Breech presentation predisposes to brachial plexus, spinal cord, and brainstem injury during birth.3,18 Autopsy studies on neonates have revealed that unilateral disruption of nerve roots, laceration of dura with epidural hemorrhage, and contusion or laceration of the spinal cord or medulla especially at the cervico-medullary junction are the most common severe injuries which result in death.19,21 Towbin22,23 studied neonates who
suffered birth trauma and found displaced fragments of cerebellum in the subarachnoid space. Their appearance is almost identical to the ectopic glial islands that we observed. The presence of cerebellar scarring confirms that cerebellum was the source of the ectopic tissue. The history of brachial plexus injury indicates that the patient’s birth was traumatic. While we have no proof that the patient had hypotonia or respiratory distress at birth, we speculate that cerebellar trauma occurred at the same time. The fact that brainstem damage was most severe at the same level suggests that there may have been a minor hematobulbia which resolved leaving only glial scar and small cavities which were the nidus to syrinx formation.24,25

Barnett has cautioned that syringomyelia as a late sequel to minor trauma is rare and must be interpreted with caution.26 Nevertheless, delayed syrinx formation apparently secondary to obstetrical trauma has been reported.27 We suggest that the trauma suffered at birth by this patient, while not sufficient to paralyze her, was severe enough to cause brachial plexus stretching and a minor cerebellar injury which resulted in arachnoid fibrosis at the foramen magnum. The latter had been shown to be an important factor in the development of syrinxes. Lack of involvement of the central canal in the medulla and upper cervical spinal cord indicates this was at least initially a “non-communicating” syringomyelia which began in the parenchyma of the medulla and eventually tunneled caudally into the central canal of the lower cervical spinal cord and rostrally into the fourth ventricle. Extension of post-traumatic syrinxes appears to be the result of fluid movements induced primarily by changes in venous volume, and large transient venous pressure changes such as those associated with coughing or straining are believed to be especially important in the enlargement of syrinxes.28-30 Obstruction to cerebrospinal fluid movement at the foramen magnum by arachnoid fibrosis serves to aggravate fluid dynamics within the syrinx and MRI has demonstrated that fluid movements within the cavities are more rapid than those in the subarachnoid space.31,32 On two occasions the patient reported herein suffered neurologic deterioration after straining or falling. Assuming that intracavitary pressure is transiently increased during such episodes, a dissecting pressure which follows the path of least resistance is created. In this and other cases of syrinxes extending from the medulla into the midbrain or diencephalon,2,3 it would appear that the low-resistance pathway is among the axon bundles of the corticospinal tracts. Only when resistance is met will the diameter of the cavity tend to enlarge. In this case, as well as the others, that site is the internal capsule where the fiber pathways begin to diverge and perhaps are not so easily separated. Within months of cavity formation the lining composed of reactive astroglia acquires a mature appearance and presumably thickens with time and continued stress.33-35 In this patient, the presence of a simple glial band along the left corticospinal tract indicates that a once patent cavity has collapsed. Whether obliteration was done to glial proliferation or loss of cavity pressure following rupture into the fourth ventricle or subarachnoid space is not clear. Because the glial bands are very narrow we would favor the latter explanation.

REFERENCES


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