CASE REPORT

NORTH BENGAL MEDICAL COLLEGE JOURNAL [2018], VOL., 4, No. 1 : 37-41

# Syringomyelia with Unusual Presentation

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**ARTICLE INFO**

***Article history:***

Received: 08 September 2017
Accepted: 14 October 2017

***Online:***

www.nbmc.ac.bd

***Keywords:***

Syrigomyelia, Syrinx, Arachnoid cyst, Arnold-Chiari.

***ABSTRACT***

Patients with syringomyelia may have diverse aetiology and experience a variety of symptoms, the commonest presentation is dissociated sensory loss affecting pain and temperature sparing touch and vibration sensation which is suspended over the nape of the neck, shoulders, and upper arms. This report describes two cases of syringomyelia with different profiles in that they presented with dominant motor deficits.

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## INTRODUCTION

Syringomyelia is a rare neurological disorder characterized by slowly developing central fluid-filled areas (cyst or syrinx) that usually involve the cervical spinal cord and expand as well as extend downwards causing progressive damage to the spinal tracts. Classic presentation is a central cord syndrome with dissociated sensory loss showing loss of pain and temperature sensation sparing touch and vibration suspended over the nape of the neck, shoulders, and upper arms in a cape distribution or is in the hands. Muscle wasting in the lower neck, shoulders, arms, and hands with asymmetric or absent reflexes reflects extension of the syrinx to the anterior horns.[1-4] More than half of all cases are associated with Arnold-Chiari type I malformations in which the cerebellar tonsil protrudes through the foramen magnum and into the cervical spinal canal.[5-7] Syringomyelia has a prevalence of 3.3 to 8.5/100,000 people with some ethnic variability.[1-4] In the United States, syringomyelia is more common in African-Americans than in Caucasians.[8] It occurs more frequently in men than in women and usually manifests in the third or fourth decade of life. Rarely, syringomyelia may develop in childhood

or late adulthood. Loss of pain sensation is the most common early neurological manifestation in syringomyelia with Chiari-1 malformation.[9]

The presenting two cases are unusual in that, they had pain and temperature sensations intact rather with features of dorsal column and pyramidal tract lesions which could otherwise suggest a diagnosis of slowly developing compressive spinal myelopathy.

## Case 1

An 18-year-old male, a university student, came to medicine outpatient department of Dhaka Community Medical College Hospital with limb weakness and gait disturbance for 3 months. In suspicion of cervical myelopathy, spinal X-ray was done and found normal.He was advised for neuroimaging. He is a resident of Sariakandi, Bogra and was later admitted in the department of Medicine, Shaheed Ziaur Rahman Medical College Hospital for the same complaints. He had no complaints of headache, visual disturbance, bladder or bowel dysfunction, fever or joint pain or history of neck trauma. His milestones of physical and mental developments were seemingly normal.

On examination, the patient was ill looking, with normal higher psychic function and speech. Cranial nerves were intact with normal optic fundi. Bulk of the muscle was reduced in both upper limbs with diminished tone, power (4 of 5) and jerks. In both lower limbs, muscle bulk was normal with hypertonia, grade 4 of 5 power and exaggerated deep tendon reflexes, extensor planter response and sustained clonus. Dorsal column sensations (sense of position and vibration) were reduced but pain and temperature sensations were intact in all 4 limbs. Romberg test was positive. Tests for cerebellar functions were normal. There was no spinal deformity. Other systemic examination revealed no abnormality.

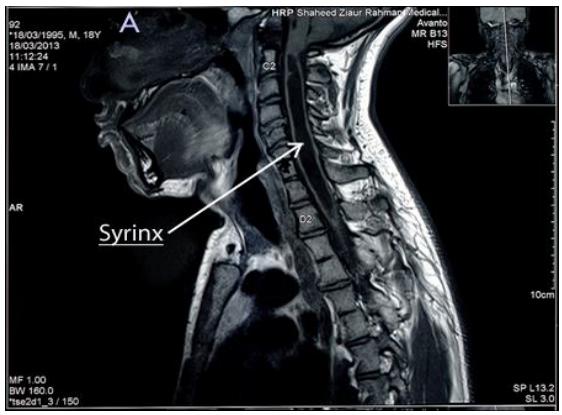


**Figure 1: MR images showing a large syrinx extending from C2 to D2**

Investigations revealed haemoglobin-12 gm/dl, total leucocyte count-8000/$mm^3$ with normal differential counts and erythrocyte sedimentation rate-15 mm in 1st hour.Urinalysis, fasting blood glucose level, renal and liver function tests and serum vitamin $B_{12}$ level were normal. MRI of cervical and dorsal spine (Figure 1**)** revealed a large syrinx extending from cervico-medullary junction to second dorsal spine without herniation of cerebellar tonsil. With these findings, the case was diagnosed as syringomyelia with unusual presentation.

## Case 2

A 35-year-old male, married, normotensive, non-diabetic, farmer hailing from Jamua, Sirajgang, presented with progressive weakness of both upper limbs as well as wasting of muscles in his upper back region for two years. There was no history of bladder or bowel dysfunction. There was no disturbance of sensation. He gave no history of tuberculosis or trauma to the head or spine. There was no history of such illness among other members of his family.

On examination, the patient was conscious and well oriented. Higher psychic functions were normal. There was no abnormality in speech. Cranial nerves were intact. Bulk of the muscle

was reduced in both upperlimbs and upper back of the trunk, which was marked around the shoulder girdle (Figure 2). There was no fasciculation. In both upper limbs, muscle tone was diminished, muscle power was grade3 of 5 and deep tendon reflexes were reduced. In the lower limbs, muscle tone, power and reflexes were normal.

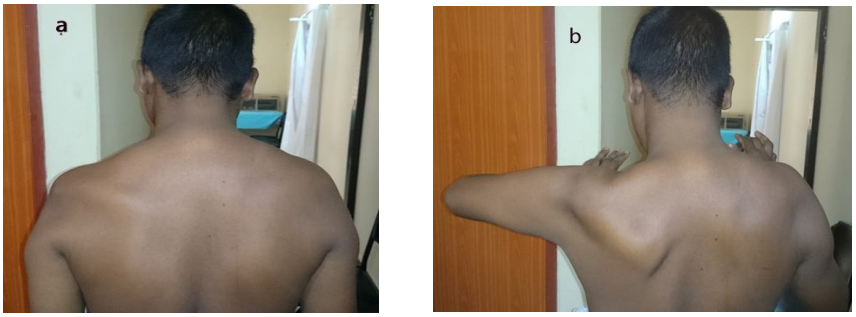


**Figure 2: Photographs (a & b) showing muscle wasting of the shoulder girdle with winging of scapula**

Plantar response was flexor in both sides. All modalities of sensations were intact. There were no signs of cerebellar lesion.

Routine investigations and biochemical values viz.urinalysis, complete blood counts, erythrocyte sedimentation rate, fasting blood glucose and serum creatinine, T3, T4 and TSH were all within the normal limits. Plain radiograph of spine was normal. MRI of cervical and dorsal spine (Figure 3) revealed a large syrinx extending from cervical to mid-dorsal region.

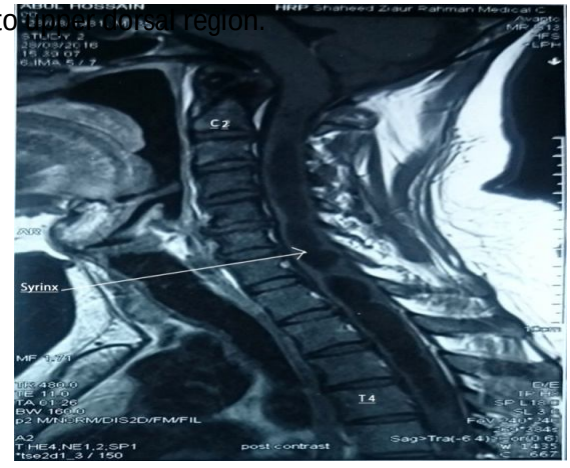


**Figure 3: MR images showing a large cervico dorsal syrinx**

## DISCUSSION

Syringomyelia is a developmental cavity (syrinx) of the cervical cord that is prone to enlarge and produce progressive myelopathy. Symptoms begin insidiously in adolescence or early adulthood, progress irregularly, and may undergo spontaneous arrest for several years. A number of pathological conditions can cause an obstruction of the normal cerebrospinal fluid (CSF) spaces. These include Chiari malfor-mation, spinal arachnoiditis (post infectious, inflamma-tory, post-irradiation or blood in subarachnoid space), scoliosis, spinal vertebrae misalignment, pathological masses (arachnoid cysts, rheumatoid arthritis pannus, occipital encephalocoele, tumours), spina bifida and others.[10,11] The reasons that cause blockage of the CSF space within the subarachnoid space can result in syrinx formation are not known. Moreover, it is unclear if syrinx fluid originates from bulk movement of CSF into the spinal cord, from bulk transmural movement of blood fluids through the spinal vasculature into the syrinx, or from a combination of both. Many young patients acquire a cervical-thoracic scoliosis. More than half of all cases are associated with Chiari type 1

malformations in which the cerebellar tonsils protrude through the foramen magnum and into the cervical spinal canal blocking the flow of CSF. Acquired cavitations of the cord in areas of necrosis are also termed syrinx cavities; these follow trauma, myelitis, necrotic spinal cord tumors, and chronic arachnoiditis due to tuberculosis and other etiologies.

The presentation is a central cord syndrome with dissociated sensory loss in cape distribution or in the hand and areflexic weakness in the upper limbs. Most cases begin asymmetrically with unilateral sensory loss in the hands that leads to injuries and burns that are not appreciated by the patient. Muscle wasting in the lower neck, shoulders, arms, and hands with asymmetric or absent reflexes in the arms reflects expansion of the cavity into the gray matter of the cord. As the cavity enlarges and further compresses the long tracts, spasticity and weakness of the legs, bladder and bowel dysfunction, and Horner's syndrome appear. Some patients develop facial numbness and sensory loss from damage to the descending tract of the trigeminal nerve (C2 level or above). In cases with Chiari malformations, cough-induced headache and neck, arm, or facial pain are reported. Extension of the syrinx into the medulla or syringobulbia causes palatal or vocal cord paralysis, dysarthria, horizontal or vertical nystagmus, episodic dizziness or vertigo, and tongue weakness with atrophy.[12]

MR imaging accurately identifies developmental and acquired syrinx cavities and their associated spinal cord enlargement. MRI images of the brain and the entire spinal cord should be obtained to delineate the full longitudinal extent of the syrinx, to assess posterior fossa structures for the Chiari malformation, and to determine whether hydrocephalus is present.[13]

Treatment of syringomyelia is not fully satisfactory. The Chiari tonsillar herniation is usually decompressed, usually by suboccipital craniectomy, upper cervical laminectomy, and placement of a dural graft. Obstruction of fourth ventricular outflow is reestablished by this procedure. If the syrinx cavity is large, some surgeons recommend direct decompression or drainage by one of a number of methods, but the added benefit of these procedures are uncertain, and morbidity is common. With Chiari malformations, shunting of hydrocephalus should generally precede any attempt to correct the syrinx. Surgery may stabilize the neurologic deficit, and some patients improve. Syringomyelia secondary to trauma or infection is treated with a decompression and drainage procedure in which a small shunt is inserted between the syrinx cavity and the subarachnoid space. Alternatively, the cavity can be fenestrated. Cases due to intramedullary spinal cord tumor are generally managed by resection of the tumor. Assessing treatment results is difficult because of the rarity of syringomyelia, variability of presentation and natural history, and the relatively short follow-up in most studies. In one study, half of cases were in clinically stable condition for several years.[14] Although an older study had suggested that 20% of patients died at an average of 47 years, mortality rates are likely lower in today's patients as a result of surgical interventions and better treatment of complications.

**Conflicts of interest:** None

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