

Analysis of Syrinx and Its Associations: A Retrospective Study

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Abstract

Background: Syrinx is described as a cavitory enlargement of the spinal cord. It is a common reason for neurosurgical referral. The aetiology of syrinx is multiple and it is associated with other brain and spinal cord conditions, while some are not associated with any other conditions. **Subjects and Methods:** We retrospectively collected data of cases diagnosed with syrinx in the department of radiodiagnosis in Rajarajeswari Medical college, Bangalore and studied the conditions associated with syrinx. **Results:** A total of 37 cases were analysed. The spinal cord levels most commonly associated with syrinx were C5, C6, T1 and L1 vertebral levels. Arnold Chiari malformation type 1 was the most common condition associated with syrinx. **Conclusion:** It was concluded from the present study that syrinx was more common in males than females with mean age of presentation of 36 years. The cervical spinal cord level was most commonly involved followed by thoracic and lumbar spinal cords. ACM type 1 was the most common associated condition. It becomes important for radiologist to look for syrinx in cases diagnosed with associated conditions, so that subtle and mild forms of syrinx are not missed and their growth or expansion in spinal cord be evaluated in follow up MRI studies.

Keywords: Syrinx, Syringomyelia, Arnold Chiari Malformation.

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Introduction

Syrinx is described as a cavitory enlargement of the spinal cord.^[1] It is a common reason for neurosurgical referral.^[2] The precise mechanism for formation, growth of syrinx is unknown and there are many theories on its formation.^[3] It is associated with many other brain and spinal cord conditions. The Objective of the present Study was to study the demographic characteristics of syrinx and presence of associated conditions. This helps to look for the presence of subtle and mild forms of syrinx when an associated condition is diagnosed. The presence of associated conditions is crucial for therapeutic planning and has a great impact on the treatment, risk of recurrence and survival. We retrospectively studied the Magnetic Resonance Imaging (MRI) reports of cases of syrinx and analysed the spectrum of associated conditions.

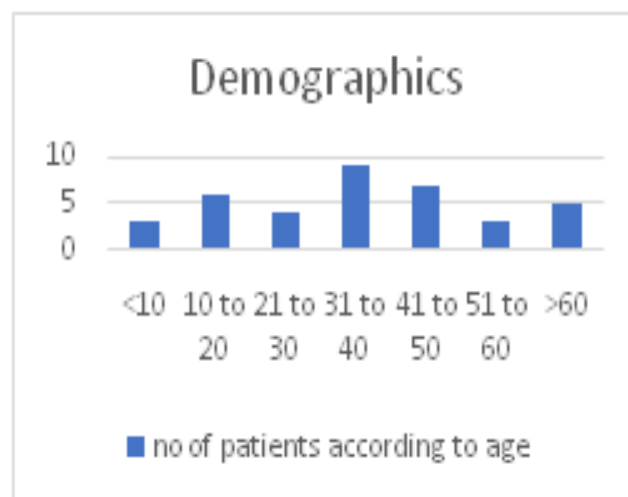
Subjects and Methods

Approval from the internal ethical committee of Rajarajeswari Medical college, Bangalore was obtained and data of all cases diagnosed to have syrinx from 2011 to 2019 in the department of radiodiagnosis was collected. A total of 37 MRI reports were reviewed and all observations were recorded on a master sheet which included patient demographics, vertebral level of spinal cord at which syrinx

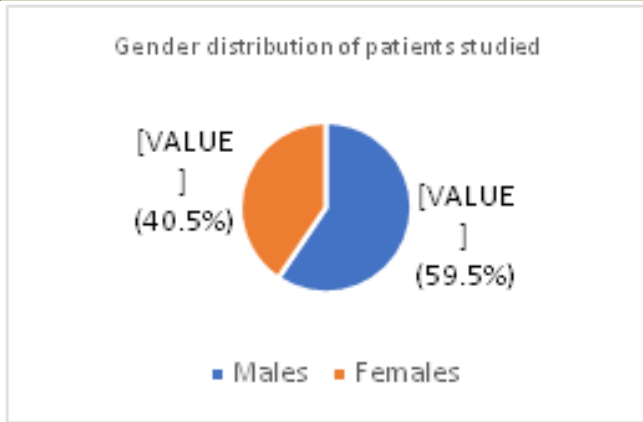
was present and associated conditions. Descriptive and inferential statistical analysis was done using Microsoft excel and SPSS software.

Results

A total of 37 cases were analysed comprising 22 males and 15 females with age ranging from 3 to 75 years and a mean age of 36 years.

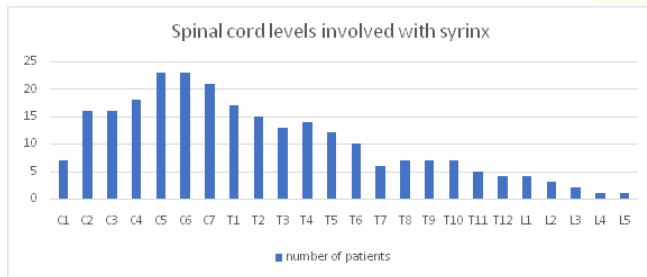


Graph 1: Age distribution of cases



Graph 2: Gender distribution of cases

The most commonly involved cervical (C)spinal cord levels were C5 and C6 both involving 62% (n=23) of cases each, the most commonly involved Thoracic(T) and Lumbar(L)spinal cord levels were T1 and L1 involving 46% (n=17) and 10.8% (n=4) of cases respectively. The lower cervical and upper thoracic spinal cord was the most commonly involved spinal cord segment, with C4 to T1 spinal cord segment involving at least 46% of cases.



Graph 3: distribution of syrinx at different spinal cord levels

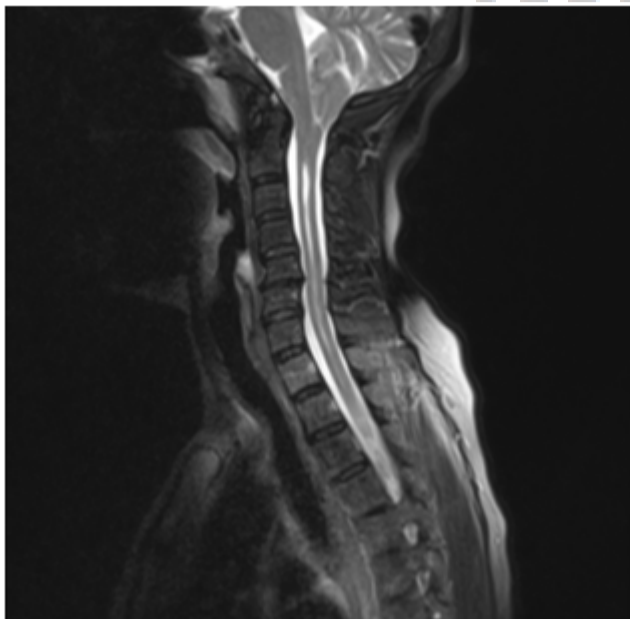


Figure 1: Saggital T2WI MRI image showing a case of cervical and thoracic spinal cord syrinx associated with ACM Type 1.

The most commonly associated condition was Arnold

Chiari malformation (ACM) Type 1 which was seen in 32% (n=12) of cases. Other commonly associated conditions were tethered cord, thoracic scoliosis (both 11%, n=4); spina bifida, congenital block vertebra (both 8%, n=3). Other associated conditions were ACM Type 2, meningomyelocele, mass in spinal cord, hypoplasticclivus, platybasia, cervical scoliosis, cervical kyphosis, lumbar scoliosis.

Table 1: Conditions associated with syrinx

| Clinical Features | Gender | | Total(n=37) |
|---------------------------|--------------|------------|-------------|
| | Female(n=15) | Male(n=22) | |
| Mass in spinal cord | 1 | 1 | 2(5.4%) |
| ACM type 1 | 6 | 6 | 12(32.4%) |
| ACM type 2 | 0 | 2 | 2(5.4%) |
| Tethered cord | 2 | 2 | 4(10.6%) |
| Meningomyelocele | 1 | 1 | 2(5.4%) |
| Spina bifida | 3 | 0 | 3(8.3%) |
| Congenital block vertebra | 1 | 2 | 3(8.1%) |
| Hypoplasticclivus | 0 | 1 | 1(2.7%) |
| Platybasia | 0 | 1 | 1(2.7%) |
| Cervical scoliosis | 1 | 0 | 1(2.7%) |
| Cervical kyphosis | 0 | 1 | 1(2.7%) |
| Thoracic scoliosis | 1 | 3 | 4(10.8%) |
| Lumbar scoliosis | 0 | 1 | 1(2.7%) |

Discussion

Syrinx is an umbrella terminology used to describe various conditions like Syringomyelia, Hydromyelia, syringobulbia, Syringopontia, Syringomesencephaly, and syringocephalus.

Syringomyelia is cavitory lesion within cord parenchyma, located adjacent to the central canal, therefore not lined by ependyma. Hydromyelia is fluid accumulation/dilatation within the central canal, therefore, lined by ependyma.

- Syringohydromyelia is the term used for either of the above, since the two may overlap and cannot be discriminated on imaging, also known as hydrosyringomyelia.^[4]
- Syringobulbia is extension of syringomyelia into the medulla oblongata.^[5]
- Syringopontia is extension of syringomyelia into the pons.^[6]
- Syringomesencephaly is extension of syringomyelia into the midbrain.^[7]
- Syringocephalus is extension of syringomyelia into the cerebrum, also known as syringoencephalomyelia.^[8]

The precise mechanism of formation, growth of syrinx is unknown and there are many theories on this. However it is associated with many other brain and spinal cord conditions like Arnold Chiari malformation (ACM), tethered cord, closed dysraphism, multiple sclerosis, trauma, spinal tumors, scoliosis, etc.^[1,9,10] Some are not associated with any of these conditions and may be considered idiopathic.^[11] Morphology of syrinx, including its

location and width, differs according to syrinx etiology.^[2] This suggests a distinct pathogenesis for syrinx with different associated conditions.^[2] When considering ACM-I and idiopathic syrinx, the combination of width greater than 5 mm and cranial extent in the cervical spine is highly specific for ACM-I associated syrinx. This finding may assist with assigning causation when evaluating a patient with both ACM-I and syringomyelia.^[2]

Syrinx is a slowly progressing condition and may extend over many years.^[12] Syringobulbia is said to have an acute course.^[13, 14] Widespread use of MRI for many other conditions has facilitated the diagnosis of subclinical syringomyelia.^[15] Symptoms depend on location of the lesion within the neuraxis. It affects sensory, motor and autonomic functions. Posttraumatic syringomyelia occurs commonly in patients with spinal cord injury (SCI).^[9, 16] Holocord syringomyelia can occur in patients with a history of SCI and present with new onset focal neurologic deficit.^[17] In patients with incidental syringomyelia without any other predisposing condition, close observation is recommended rather than surgery.

MRI is the imaging modality of choice and syrinx characteristically follows CSF signal characteristics on all sequences, that is hypointense on T1WI, hyperintense on T2WI.^[18, 19] Both sagittal and axial images have to be obtained and the entire craniocaudal extent of the syrinx should be evaluated.^[20] Unenhanced MRI that includes sagittal and axial T2-weighted imaging sequences appears to have a high sensitivity and high negative predictive value in the evaluation for a syrinx associated mass, and contrast material may not be required to assess all cases of syrinx.^[21]

Conclusion

It was concluded from the present study that syrinx was more common in males than females with mean age of presentation of 36 years. The cervical spinal cord level was most commonly involved followed by thoracic and lumbar spinal cords. ACM type 1 was the most common associated condition. It becomes important for radiologist to look for syrinx in cases diagnosed with associated conditions, so that subtle and mild forms of syrinx are not missed and their growth or expansion in spinal cord be evaluated in follow up MRI studies.

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