

Unmasking the Mystery of A Hidden Syrinx: Case Report of Progressive Paralysis in A 20-Year-Old Male with Intact Pain and Temperature Sensations

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ABSTRACT

Syringomyelia is a progressive neurological disorder in which a fluid-filled cyst known as syrinx forms within the spinal cord. The case of a 20-year of adult is presented with lower limb stiffness and difficulty in walking for last 3 years. For the last 1 year, he also developed stiffness of upper limbs. Upon examination, he had a spastic gait with no ataxia. There was muscle wasting of both upper and lower limbs, with fasciculation. Tone was increased with reduced power and hyperreflexia bilaterally in all four limbs with positive ankle clonus and upgoing plantars bilaterally. Sensory system, cerebellum and higher motor functions were intact. MRI scan revealed an abnormal intramedullary multifocal T2WI hyper intense signal in cervical and thoracic spinal cord consistent with syrinx and he was diagnosed with Syringomyelia. This case is an uncommon presentation of Syringomyelia presenting with predominantly upper motor neuron lesion without sensory involvement.

Keywords: MRI Scan, Syringomyelia, Syrinx in Spinal Cord.

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INTRODUCTION:

Cerebrospinal fluid (CSF) normally surrounds and protects the brain and spinal cord. However the CSF may build up within the tissue of the spinal cord to expand the central canal forming a syrinx. Syringomyelia is a progressive neurological disorder in which a fluid-filled cyst known as syrinx forms within the spinal cord.¹ When the syrinx affects

the brain stem, it is called syringobulbia.¹ The syrinx can get big enough to damage the spinal cord and cause compression injury of various spinal nerve tracks. Clinical features of syringomyelia are various depending on the site, size and spinal extension of the syrinx.² Clinical features are usually slow to develop, progress with time and can affect one or both sides of the body. Symptoms may include loss of pain and temperature sensations, chronic pain, stiffness and motor weakness of upper limbs, headaches, loss of balance, urinary and fecal incontinence, sexual dysfunction and/or scoliosis of the spine.^{1,2}

There are two main forms of syringomyelia i.e; Congenital and Acquired. Congenital syringomyelia is usually caused by Arnold-Chiari malformation, an abnormal condition in which brain tissue extends through the foramen magnum into the spinal canal obstructing the flow of CSF to result in syrinx formation in the cervical spine.^{3,4} Symptoms commonly start between ages 25 and 40 years. People with congenital syringomyelia may also have hydrocephalus, hence it is also called communicating syringomyelia. Acquired syringomyelia (primary spinal syringomyelia or non-communicating syringomyelia) may be caused by spinal cord injuries, hemorrhage, spinal cord tumors, meningitis, arachnoiditis and tethered cord syndrome or it may be idiopathic.^{3,5}

CASE:

The case of a previously-healthy 20-years adult presented with insidious onset, gradually worsening lower limb stiffness and difficulty in walking for last 3 years. He was able to walk but required assistance. For the last 1 year, he also developed insidious onset, gradually worsening stiffness of

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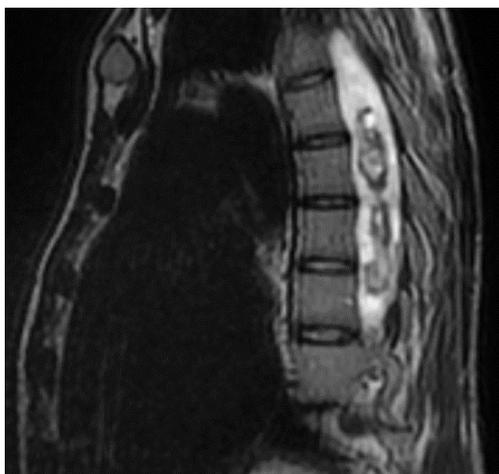
upper limbs causing him difficulties to do vocational, avocational and self-care activities. He had stopped working as a mechanic 3 months previously due to his illness. Upon probing; there was no history of numbness or tingling of limbs, spinal trauma, fits, psychiatric symptoms, weight loss, night sweats, urinary or fecal incontinence, altered bowel habits, joint pains or any palpable lumps. He was unmarried and denied sexual contact. He did not smoke or use illicit drugs. Although his parents were first cousins, there was no family history of any similar disorder.

Upon examination, the patient had a spastic gait with no

Figure 1: Abnormal intramedullary multifocal T2WI hyperintense signals seen in the cervical spinal cord on MRI scan



Figure 2: Abnormal intramedullary multifocal T2WI hyperintense signals seen in the thoracic spinal cord on MRI scan



ataxia. There was muscle wasting of both upper and lower limbs, with fasciculation present. In the upper limbs, tone was increased, power 4/5 with Grade III hyperreflexia bilaterally. In lower limbs, tone was increased, power 3/5 with Grade IV hyperreflexia, positive ankle clonus and up going plantars bilaterally. There was no loss of sensation of lateral (pain, temperature) or posterior (proprioception, vibration) spinothalamic tracts. Cerebellum and higher motor functions were intact. There was no tenderness, gibbus or deformity of the spinal vertebrae on back examination. On MRI scan, abnormal intramedullary multifocal T2WI hyperintense signals were seen in cervical and thoracic spinal cord consistent with syringomyelia as shown in Figures 1 and 2. Based on history, examination and radiographic findings, he was diagnosed with Idiopathic Acquired Syringomyelia. This case is an uncommon presentation of Syringomyelia presenting with predominantly upper motor neuron lesion without sensory involvement.

DISCUSSION:

Motor weakness, muscle wasting, and decreased reflexes may develop in the upper extremities due to damage to the anterior horn cells and the corticospinal tract in the cervical spinal cord.⁶ Involvement of the lower extremities is usually rare but can occur if the syrinx extends down the spinal cord results in spasticity, hyperreflexia, and positive Babinski sign.⁷ This case had muscle wasting of both upper and lower limbs, increased tone with reduced power and hyperreflexia bilaterally in all four limbs with positive ankle clonus and upgoing plantars bilaterally. MRI scan discovered the syrinx extending from the cervical to thoracic spine which helps to explain his examination findings. Loss of pain and temperature sensation, bilaterally and symmetrically in the upper limbs, upper trunk and neck, is usually the first symptom of syringomyelia and is due to the involvement of the lateral spinothalamic tract.^{6,7} Proprioception, vibration sense, and fine touch are preserved, as these sensory pathways are located more dorsally in the spinal cord and usually remain preserved in syringomyelia.⁶ Autonomic symptoms such as sweating abnormalities, vasomotor changes, bladder and bowel dysfunction may occur due to involvement of the sympathetic and parasympathetic pathways within the spinal cord.⁷ However, this patient did not have sensory loss or autonomic abnormalities.

Magnetic resonance imaging (MRI) of the spinal cord is the most reliable way to diagnose syringomyelia.⁸ MRI not only helps to determine the presence, size and extent of syrinx in the spinal cord but also aids in ruling out other causes such as spinal cord injury/compression, tumor, meningitis, hemorrhage and Arnold-Chiari malformation.^{7,8} Treatment for syringomyelia depends on the severity and progression of symptoms.⁹ In asymptomatic and mild cases, a conservative management plan focused on physiotherapy, avoidance of activities that cause strain on spine and close observation is usually employed.¹⁰ In more severe symptomatic or

progressive disease, the solution is surgery aimed to eliminate the syrinx and prevent further spinal cord injury.^{9,10} There are two general forms of surgical treatment: restoration of normal CSF flow around the spinal cord, and directly draining the syrinx depending on the symptoms and severity of the disease.¹¹

Osama et al.¹² reported a case of C6-C7 level syrinx presenting as neck pain, radiating to his right arm, associated with paresthesia involving Index and Middle finger and a positive spurling test with postural deviation and associated disability. The patient received physical therapy management consisting of pain management, cervical traction, joint mobilization and soft tissue manual therapy aimed at postural and biomechanical correction in combination with medications resulting in marked improvement.¹² Butt et al.¹³ reported 2 cases of syringomyelia, first was the result of craniocervical meningioma and the second was due to gunshot wound leading to spinal injury. Both cases required surgical intervention and reported improvements afterwards.¹³ Ihsanullah et al.¹⁴ assessed the post-operative outcome after Posterior Fossa Decompression (PFD) with duraplasty in 28 patients with Chiari-1 malformations to report good outcome in 22 patients and fair outcome in 6 cases. Furthermore, syringomyelia cases are recovered well with syringe-subarachnoid shunt.¹⁴

In conclusion, bilateral upper and lower limb weakness of upper motor neuron type is a rare presentation of syringomyelia and it can be challenging to diagnose. The presence of a syrinx within the spinal cord can cause damage to different sensory and motor pathways leading to a wide range of symptoms. This patient has uncommon presentation of Syringomyelia presenting with predominantly upper motor neuron lesion without sensory involvement. The diagnosis of syringomyelia was confirmed by MRI scan which showed the characteristic features of the disease.

Authors Contribution:

Muhammad Sohail Ajmal Ghoauri: Conception and design, data collection, assembly and patient assessment, manuscript writing

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REFERENCES:

1. Kim J, Kim CH, Jahng TA, Chung CK. Clinical course of incidental syringomyelia without predisposing pathologies. *J Clin Neurosci.* 2012;19(5):665-8. doi: 10.1016/j.jocn.2011.07.050.

2. Sixt C, Riether F, Will BE, Tatagiba MS, Roser F. Evaluation of quality of life parameters in patients who have syringomyelia. *J Clin Neurosci.* 2009;16(12):1599-603. doi: 10.1016/j.jocn.2009.04.019.
3. Lucchetta M, Cagnin A, Calderone M, Manara R, Rotilio A, Briani C. Syringomyelia associated with Chiari I malformation. *Neurol Sci.* 2009;30(6):525-6. doi: 10.1007/s10072-009-0133-8.
4. Nacir B, Arslan Cebeci S, Cetinkaya E, Karagoz A, Erdem HR. Neuropathic arthropathy progressing with multiple joint involvement in the upper extremity due to syringomyelia and type I Arnold-Chiari malformation. *Rheumatol Int.* 2010;30(7):979-83. doi: 10.1007/s00296-009-1013-5.
5. Nakanishi K, Uchiyama T, Nakano N, Fukawa N, Yamada K, Yabuuchi T, Kato A. Spinal syringomyelia following subarachnoid hemorrhage. *J Clin Neurosci.* 2012 ;19(4):594-7. doi: 10.1016/j.jocn.2011.07.035.
6. Wu J, Ji S, Niu P, Zhang B, Shao D, Li Y, et al. Knowledge mapping of syringomyelia from 2003 to 2022: A bibliometric analysis. *J Clin Neurosci.* 2023;110:63-70. doi: 10.1016/j.jocn.2023.01.004.
7. Holste KG, Muraszko KM, Maher CO. Epidemiology of Chiari I Malformation and Syringomyelia. *Neurosurg Clin N Am.* 2023;34(1):9-15. doi: 10.1016/j.nec.2022.08.001.
8. Spennato P, Vitulli F, Tafuto R, Imperato A, Mirone G, Cinalli G. Fourth ventricle to spinal subarachnoid space stenting in pediatric patients with refractory syringomyelia: case series and systematic review. *Neurosurg Rev.* 2023;46(1):67. doi: 10.1007/s10143-023-01972-y.
9. Sato S, Kakizaki T, Kondo K, Kanno C, Maeda Y, Ando R, et al. Arnold Chiari malformation classified as Chiari 1.5 malformation in a Japanese black calf. *J Vet Med Sci.* 2023. doi: 10.1292/jvms.22-0463.
10. Hersh AM, Lubelski D, Theodore N, Sciubba DM, Jallo G, Shimony N. Approaches to Incidental Intradural Tumors of the Spine in the Pediatric Population. *Pediatr Neurosurg.* 2023. doi: 10.1159/000530286.
11. Lu C, Guan J, Ding C, Wang X, Wang Z, Chen Z, et al. Post-traumatic syringomyelia resolution following surgical treatment: the moniliform syrinx with a better prognosis. *Acta Neurol Belg.* 2023. doi: 10.1007/s13760-023-02233-x.
12. Osama M, Yaqoob F. Cervical syringomyelia; conservative physical therapy management of a patient: a case report. *Professional Med J.* 2017;24(4):627-632. DOI: 10.17957/TPMJ/17.3817.
13. Butt B, Zunair SA, Rehman A, Latif A, Awan NR, Anwar K, et al. Post-Traumatic Syringomyelia: A Case Series. *Pak. J. of Neurol. Surg.* 2021; 25(4): 569-578. DOI:10.36552/pjns.v25i4.626.
14. Ihsanullah, Khanzada K, Alam M, Hameedullah. Surgical Outcome of Chiari Malformation Type One in Adults. *Pak. J. of Neurol. Surg.* 2019; 23(1): 14-18.