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# Syringomyelia due to cervical spondylosis with an atypical sensory deficit – A rare and unusual presentation

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## Abstract

Syringomyelia is a rare neurological disorder that is progressive and characterized by the formation of a fluid-filled cavity in the spinal cord. Causes of this condition may be congenital or acquired, however, the latter can rarely occur due to cervical spondylosis. Here we report a presentation of syringomyelia in a 59-year-old man with gradual onset weakness and muscle wasting along with atypical sensory deficit of affected temperature but intact pain modalities in upper limbs. The MRI of the cervical spine confirmed the presence of a syrinx from C2 to D2/D3 levels associated with degenerative cervical spinal disease with canal stenosis. Surgery remains the primary treatment modality for symptomatic disease although this patient opted for complementary medicine. This case underscores the importance of considering degenerative spinal disease as an aetiology for syringomyelia and to embark on early diagnosis and individualized management strategies to prevent irreversible neurological damage.

**Key words:** syringomyelia, acquired syringomyelia, cervical spondylosis

## Introduction

Syringomyelia is an uncommon, progressive neurological disorder due to the formation of a fluid-filled cavity (syrinx) within the spinal cord. The cavity can extend to involve the brain stem when so is termed syringobulbia. The presentation of the disease depends on the extent of the syrinx. Syringomyelia occurs most commonly due to congenital causes, such as Chiari malformations.[1] Acquired causes can arise due to post-infective, post-inflammatory post-traumatic, and idiopathic causes as well as cervical spondylosis.[2] We report a presentation of a male with syringomyelia due to significant degenerative changes in the cervical spine.

## Case presentation

A 69-year-old male with well-controlled hypertension for two years duration presented with weakness and wasting of bilateral hands for two years. The weakness was of gradual onset associated with distal wasting initially of the left upper limb. Thereafter it manifested in the right upper limb however with an asymmetry of left predominance. Although there was a burning sensation in the hands, there was no pain, or recurrent trauma or wounds. He reported some intermittent episodes of neck pain associated with unsteadiness while mobilising. The weakness and wasting manifested in the left leg after 6 months and then gradually involved the right leg. The patient denied constitutional symptoms of anorexia, weight loss or fever. There were no respiratory tract symptoms, and his bladder, bowel and sexual functions were preserved. The patient denied a history of high-impact spinal trauma or significant infections. His past medical history was insignificant and there were no familial diseases including neurological disease. He was a retired mason and denied significant exposure to occupational or recreational toxic agents. He was partially dependent with a Barthel index of 55/100 requiring help, particularly in feeding, bathing and grooming. Furthermore, he was frail with a Fried phenotype of 4/5 with positive responses to self-reported exhaustion, slow walking speed, poor handgrip and low physical activity status.

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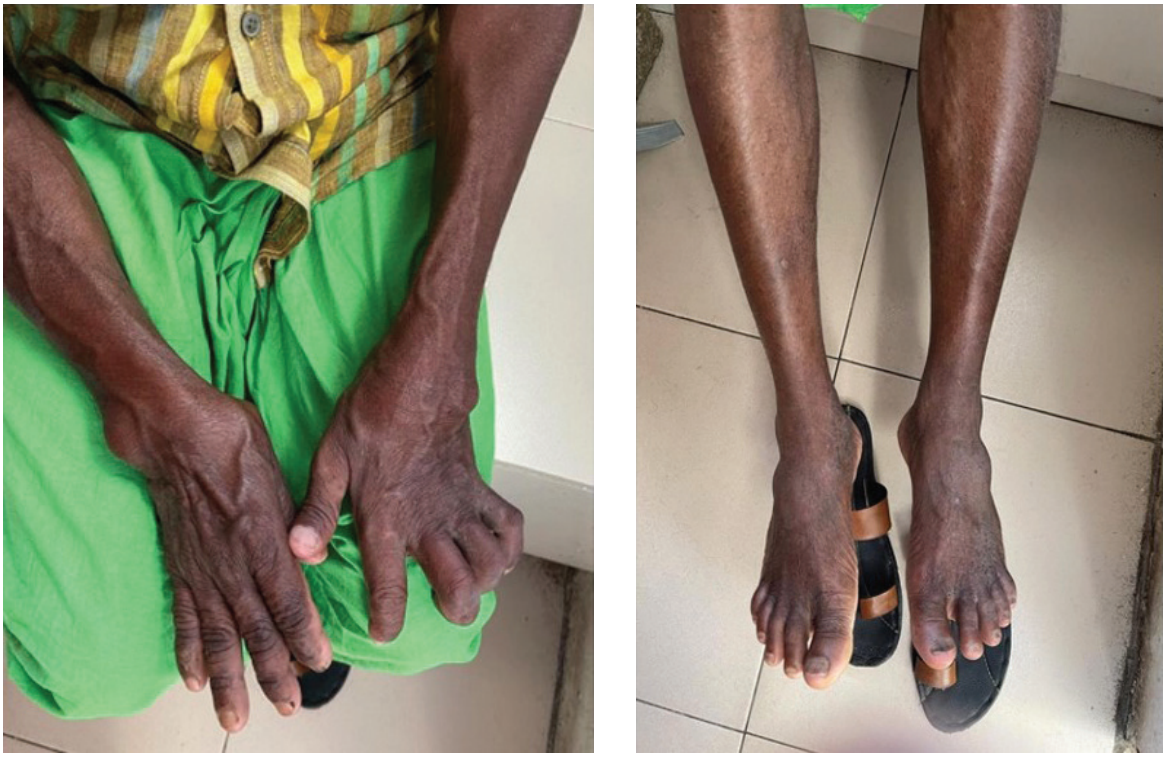
He was thin-built with a body mass index of 19 kg/m$^2$. There were no dysmorphic features. The spine was normal in curvatures without neural tube defects, kyphoscoliosis or tenderness. His upper limb examination revealed a claw hand on the left side and a wasting of small muscles (Figure 1a). There were fasciculations and bilateral asymmetric (left more than right) distal predominant wasting involving small muscles of the hands. Bilateral wrist joint swelling along with a 'loose bag of bones' appearance upon passive movements of the hand joints revealed the presence of Charcot's joints. The tone was normal with distal power of 2/5 and proximal power of 3/5 on the left and 4/5 on the right side. All upper limb reflexes were diminished. There was a dissociative sensory loss in a 'cape-like distribution' with reduced temperature sensation but intact light touch, pinprick, vibration and proprioception. His lower limb examination revealed no muscle wasting but had increased tone bilaterally (Figure 1b). Bilateral knee and ankle reflexes were exaggerated with extensor plantar response without sensory impairment. There were no cerebellar signs. There were no hypopigmented anaesthetic patches or palpable nerves. The blood pressure was 140/80 mmHg with no orthostatic hypotension while the pulse rate was 80/min and regular. Respiratory, cardiovascular and abdominal examinations were unremarkable and there was no lymphadenopathy.

The complete blood count was normal with a haemoglobin of 13.4 g/dL and a white cell count of 8.1x10$^9$/L. The erythrocyte sedimentation rate was 15mm in the first hour with a c-reactive protein of 5 mg/L. The rest of the biochemical tests were also unremarkable. His VDRL and HIV antigen-antibody status was negative. The cerebrospinal fluid analysis yielded a clear appearance with a normal protein content of 38 mg/dl and a normal glucose level of 80mg/dl. Furthermore, there were unremarkable red cell counts and differential white cell counts while the gram stain, bacterial culture and virology were negative. His chest and cervical spine radiography were normal. Nerve conduction revealed reduced amplitude of compound motor action potentials in upper limbs with normal conduction velocity of motor nerves. Chronic denervation changes with large motor unit potentials were observed in C5 to T1 myotomes bilaterally. The sensory nerves had normal conduction velocities and amplitudes. He underwent magnetic resonance imaging (MRI) of the cervical spine which demonstrated syrinx formation extending from the C2 level to the D2 level. There were posterior disc osteophyte complexes at C3/4, C4/5, C5/6 and C6/7, causing canal stenosis and cord compression with no evidence of syringobulbia. (Figure 2a, 2b) These findings were suggestive of acquired syringomyelia.



A B

**Figure 1. a) Bilateral wasting and clawing of hands. b) Normal lower limbs on inspection.**

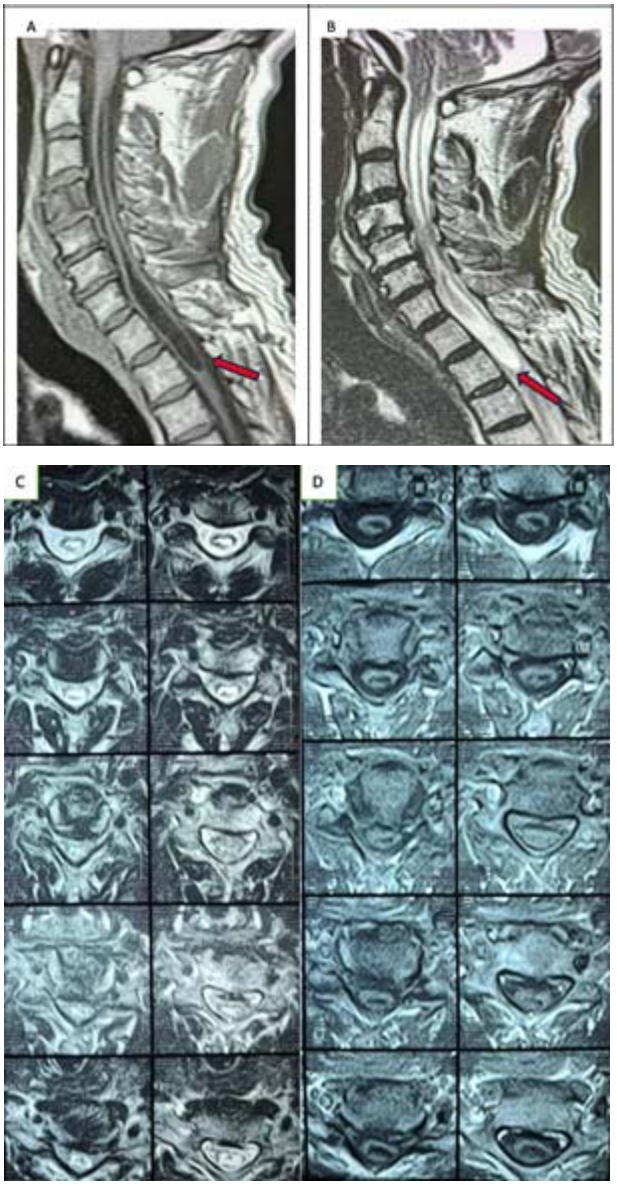



**Figure 2. A) T1 and B) T2 sagittal MRI images demonstrating syrinx formation (red arrows), C) T1 and D) T2 axial MRI images.**

The patient was referred for neurosurgical consultation with follow-up at the medical clinic. However, the patient abandoned all forms of allopathic medication in search of complementary medicine by choice. He was lost to follow up.

## Discussion

The prevalence of syringomyelia is approximately 8 per 100,000 with a male predominance.[3] Congenital aetiology of syringomyelia includes Chiari malformation type I, Klippel-Feil syndrome and tethered spinal cord.[4] Acquired presentation can arise due to post-infectious causes such as tuberculosis, and due to post-inflammatory conditions such as transverse myelitis and multiple sclerosis. Post-traumatic syringomyelia can occur in spinal cord injury patients.[5] Spinal neoplasms such as intramedullary meningiomas, ependymomas and haemangioblastoma can also result in the acquired disease.[6]

Syringomyelia as in our patient could also be rarely caused by cervical spondylosis, a common spinal degenerative condition.[7] This infrequent association can be primarily attributed to the obstruction of cerebrospinal fluid (CSF) pathways. Disc herniation, local kyphosis, osteophyte formation as well as ligamentum flavum hypertrophy can result in the narrowing of the cervical subarachnoid space. Ischaemia and microtrauma to the spinal cord following these give rise to the formation of a syrinx at the same spinal level due to myelomalacia and central canal fluid egressions.[8,9] Obstruction to CSF flow dynamics can also result in syrinx formation due to stenosis or subarachnoid adhesions.[10] The subarachnoid obstruction theory descries increased CSF pressure above the blockage causing vascular spasms, disruption of blood brain barrier causing cystic dilatation of the spinal cord.[11] Similarly fluid can dissect at the weak areas of the spinal cord. Furthermore the static and dynamic compression of flexion-extension movement of the neck can further progress spinal cord compression by ischaemic damage and fluid accumulation.[11] The presyrinx concept of predisposed anatomical changes is also a causation of syringomyelia.[11] A case series demonstrated that the most frequent levels to encounter such occurrences were C5-6, C6-7, and C4-5 and that surgical intervention often resulted in some degree of resolution of the syrinx.[12] Cervical spondylotic myelopathy is also a rarely associated occurrence with syringomyelia presenting with neck pain, limb weakness and neuroarthropathy of the hip.[9,13]

Most patients present with progressive central cord deficits with prominent pain syndrome that is exacerbated by movement.[8] The classic presentation is loss of pain and temperature sensation with preserved light touch, vibration and proprioception (dissociative sensory loss).[14] This phenomenon is due to the syringeal disruption of the spinothalamic tract which carries pain and temperature sensation with intact posterior tracts that transmit the rest of the sensory modalities. This patient only had a loss of temperature sensation with preservation of pain, light touch, vibration and proprioception. Initially, amyotrophic lateral sclerosis was considered as the diagnosis as sensory symptoms were minimal. However, the presence of Charcot's joints in the upper limbs prompted us to evaluate the presence of a syrinx. Selective loss of one sensory modality over the other

can be attributed to anatomical variations as well as specific characteristics of the syrinx such as asymmetrical growth or impact on certain fibres within the tract. Although the fibres for pain and temperature are closely aligned, their exact arrangement can vary slightly, leading to differential vulnerability. Furthermore, fibres that transmit temperature are often located in the central part of the spinal cord where the syrinx generates.[15] Bray et al. described a similar presentation of selective loss of temperature.[16] Another patient with syringomyelia was reported by Ghoauri et al with intact both modalities.[15] An atypical presentation of syringomyelia with intact pain and temperature modalities but with affected proprioception and deep pain was reported by Yohei et al. which was explained by the presence of ossified yellow ligament and arachnoid webs causing dorsal compression of the spinal cord.[17]

Risk factors for neurological progression are arachnoiditis, bony deformities, cord compression and narrowed spinal canal.[14] The diagnostic workup includes neuroimaging with Magnetic Resonance Imaging (MRI) which is the most sensitive investigation to visualize syrinx. MRI will show the size, extent and location of the syrinx and it will provide an aetiological diagnosis such as the presence of Chiari malformation, spinal tumours and enhanced leptomeninges in infection.[6]

There is no medical treatment for syringomyelia. Most patients will benefit from surgery but not all patients need surgical interventions. Mild symptomatic cases could be managed conservatively as some might resolve spontaneously.[18-20] However, for symptomatic patients surgical therapy should be opted to restore the physiological flow of CSF. In presentations with significant instability of the spine, surgical stabilization is required to prevent further spinal cord damage which is shown to improve outcomes.[8] There are several surgical methods such as suboccipital and cervical decompression, laminectomy and syringotomy, shunts, terminal ventriculostomy and neuroendoscopic surgery have been used in surgical management.[21-23] Rehabilitation with physiotherapy and occupation therapy is essential to preserve neurologic function. The disease must be detected at an early stage to prevent progressive residual effects. Patients such as this would have unfortunate outcomes including increased requirement of dependence and poor quality of life. There are ongoing debates about the best clinical practices and treatment modalities.[5] Prognosis depends on the aetiology, location, extension and neurologic deficit. The most debilitating complication of syringomyelia is progressive myelopathy which can cause quadriplegia, bladder-bowel-sexual dysfunction and decubitus ulcers.[5]

## Conclusion

Syringomyelia is a rare neurological disease which causes progressive myelopathy with serious complications particularly challenging when presenting degenerative spinal disease. This case highlights cervical spondylosis as a rare cause of acquired syrinx, describing the possible pathophysiology and highlighting the importance detecting potential causes such as cervical spondylosis, canal stenosis and cervical spondylotic myelopathy. Furthermore patients can present with atypical sensory deficits as described. Early diagnosis and treatment are paramount to prevent permanent neurological disability. There is no definitive medical therapy but surgical options may provide relief for symptomatic patients although some may need a conservative approach.

## Author declaration

### Consent for publication

Informed consent should be taken from the patient.

### Conflict of interests

The authors declare that they do not have any conflict of interests.

### Author contributions

SS and LW were directly involved in the patient care, conceptualizing, writing and review of the manuscript.

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