Syringomyelia secondary to cervical spondylosis: Case report and review of literature

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ABSTRACT

Syringomyelia secondary to cervical spondylosis is a rare entity to encounter in clinical practice. We discuss the case of a 53-year-old lady who presented with a syringomyelic syndrome and was found to have cervical spondylosis on imaging. Cine-MRI revealed an obstruction of cerebrospinal fluid (CSF) flow in the cervical spinal subarachnoid space. Decompression of the same led to clinical and radiological improvement. There is a potential causal association between cervical spondylosis and syringomyelia. MRI CSF flow studies may help in deciding the course of treatment in such cases. A subset of patients with cervical spondylosis and concurrent spinal cord signal intensity changes may show reversal of the same following intervention.

Key words: Cervical spondylosis, cine-magnetic resonance imaging, laminectomy, syringomyelia

Introduction

Syringomyelia has commonly been associated with the Chiari 1 malformation and is believed to be due to disequilibrium between the cranial and spinal subarachnoid spaces secondary to tonsilar impaction at the foramen magnum. This condition may also occur secondary to cervical spinal cord trauma, tubercular arachnoiditis and as an idiopathic entity. Cervical spondylosis is classically seen as a disease which causes extradural spinal cord compression. We present a case in which a patient with cervical spondylosis presented with a near holocord syringomyelia.

Case Report

A 53-year-old housewife presented with history of weakness and wasting of the right forearm and hand since 1 year. She also complained of significant almost disabling parasthesias in all four limbs involving both hands and feet since 6 months. In addition, she noted impaired pain and temperature sensation in the right upper limb. Spinal examination was normal. Neurologically, she had weakness and wasting of the small muscles of the right hand, with normal tone. Deep tendon reflexes were diminished in both upper limbs and normal in the lower limbs. Plantar reflexes were bilaterally flexor. She had sensory loss to pain and temperature in bilateral C5 and C6 dermatomes.

Imaging

The patient was evaluated with an MRI of the craniocervical junction and the whole spine, which revealed spondylotic changes in the cervical spine predominantly from C4-C6. A holocord syrinx, hypointense on T1 and hyperintense on T2 was seen. There was no evidence of tonsilar descent and the cistern magna was well seen [Figure 1a-d].

Cine MRI was performed to document CSF flow which showed obstruction of CSF flow in the spinal subarachnoid space opposite C4-C6 and free flow of CSF both anterior and posterior to the spinal cord at the foramen magnum [Figure 2a and b].

Dynamic flexion-extension x-rays of the cervical spine did not show any instability. Normal lordosis was maintained.

Access this article online

Quick Response Code:
Website: www.ruralneuropractice.com
DOI: 10.4103/0976-3147.145215

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Management
The patient underwent a decompressive laminectomy from C3-C6 and recovered uneventfully following surgery. She was followed up 6 weeks, 3 months and 6 months following surgery and reported significant reduction in the parasthesias. An MRI was performed 3 months following surgery which showed almost complete resolution of the syrinx with small residual signal changes opposite C4 [Figure 3a and b].

Discussion
The association of cervical spondylosis with syrinx is an extremely rare entity with very few previous reports in literature. Lucci, et al. in a description of three cases of neurogenic osteopathy secondary to a syringomyelia found that one of their cases had an associated cervical compressive element at C4.[2] Kimura et al., in 2004, reported a patient with radicular symptoms and a syringomyelic syndrome who was found to have cervical spondylosis with focal C4-5 instability who underwent decompression and stabilization. The patient improved following surgery and a post operative MRI 2 months following surgery showed marked resolution of the syrinx.[3] Rebai et al. reported a 70-year-old patient with features of
syringomyelic syndrome who had evidence of cervical cord compression secondary to multiple cervical disc bulges and an associated syringomyelobulbia. The patients symptoms and the syrinx resolved following extensive decompressive laminectomy, similar to the case reported herein.\[^4\] Kameyama and colleagues described cervical cord changed in nine patients of ossified posterior longitudinal ligament and had one case with a cervical syrinx with significant compression, who improved with immobilization of the neck.\[^5\]

Kaar \etal reported a patient with cervical spondylotic radiculomyelopathy with significant cord compression at C4-6, who underwent anterior decompression and fusion. The patient improved symptomatically but post operative MRI did not show any significant alteration in cord changes.\[^6\]

In 2006, Butteriss, \etal reported another patient of cervical spondylosis who had presented with spastic quadriplegia and was found to have severe cervical spondylosis with compression at C5/6 and C6/7 with an additional syrinx from C7 to D6. Their patient, however, declined surgical intervention.\[^7\]

In a paper on seven cases of syringomyelia without Chiari malformation, Lee \etal noted arachnoiditis, post laminectomy kyphosis, hydrocephalus and focal compressive lesions at D7 and L1 to be associated

| Table 1: Reported cases of syrinx secondary to spinal cord compression |
|-----------------------------|------------------|-----------------|------------------|------------------|------------------|------------------|
| **Author/year**               | **Age/sex** | **Presentation** | **Imaging** | **Treatment** | **Follow-up symptoms** | **Follow-up imaging** |
| Lucci, 1981\[^2\] | 56/M | Syringomyelic, arthropathy | C4 osteophyte with syrinx | - | - | - |
| Kaneyama, 1995\[^5\] | 59/M | Myelopathy | C3-4, C6-7 compression. Syrinx at C1-3/4 | Immobilization | Upper limb symptoms resolved | - |
| Kaar, 1996\[^6\] | 71/F | Syringomyelic syndrome, L>R | C3-4 instability Syrinx - C1-D1 | Posterior decompression, fusion C3/4 | Not specified, improved | No change in syrinx |
| Rebai, 2002\[^4\] | 70/M | Syringomyelic syndrome | Multiple cervical discs syringomyelobulbia | Decompressive laminectomy | 6 months improved | Resolution |
| Lee, 2002\[^8\] | 18/M | Ataxia, L weakness | Hydrocephalus, syringobulbia holocord syrinx | VP/SP shunt, FMD | No improvement | No change |
| 28/M | Paraparesis | Post laminectomy kyphosis - C4/5, syrinx C2-T2 | Anterior fusion C4-6, posterior fusion C3-5 | Neurologically Intact | Reduced to C2-C6 |
| 55/M | Left lower limb pain and weakness | Post traumatic pseudomeningeal cyst D6-8 syrinx - thoracic | Removal of cyst, augmentation duraplasty | Improved | No change |
| 38/M | Paraparesis, voiding dysfunction | Syrinx D6-D10, D12-L2, post traumatic arachnoiditis | Laminectomy+SS Shunt, with revision to SP | Mild | Resolution |
| 47/M | Left lower limb pain and weakness | Syrinx D4-D9 TB arachnoiditis | D7-10 Laminectomy, adhesiolysis | Mild | No change |
| 44/M | Right hemiparesis | Hydrocephalus, holocord syrinx | VP Shunt | Improved | Resolution |
| 48/F | Right parasthesias, right lower limb paresis | L1 compression fracture, holocord syrinx | D12-L2 decompression and fusion, SS shunt | Improved | No change |
| Kimura, 2004\[^3\] | 64/F | Cervical radiculopathy with syringomyelic syndrome | C4/5 C6/7 compression syrinx - C2-D3 | C4/5 C6/7 disectomy+cage | 2 months improved | Resolution |
| Butteriss, 2006\[^7\] | 70/M | Cervical myelopathy | C5/6 syrinx - C7-D6 | Refused treatment | - | - |
| Landi, 2013\[^9\] | 66/F | Cervical myelopathy | C3-T1 syrinx, disc bulges on extension C34,45,56,67,C7-D1 | 4 level laminectomy and posterior fusion | Mild improvement | Worsening of syrinx |
| Present | 53/F | Syringomyelic syndrome | C4-6 compression, holocord syrinx | C3-4 laminectomy | 6 months improved | Resolution |

VP: Ventriculoperitoneal, SP: Syringoperitoneal, SS: Syringosubarachnoid, FMD: Foramen magnum decompression
An extensive review by Klekamp outlines these theories. Leading to transmural movement of fluid, a dissociation of pressure above and below the block effect secondary to a local block in CSF flow causing myelomalacia and caviation, of tracts, syringomyelia include ischemia causing degeneration and outcome of patients with these conditions are shown in Table 1.

The theories of genesis of syrinx secondary to cervical spondylosis include ischemia causing degeneration of tracts, microtrauma at the site of compression causing myelomalacia and caviation, a sloshing effect secondary to a local block in CSF flow and dissociation of pressure above and below the block leading to transmural movement of fluid. An extensive review by Klekamp outlines these theories. The most recent theory is the one where the syrinx itself is formed by extracellular fluid movement into the spinal cord parenchyma due to a dissociation of intraparenchymal and subarachnoid pulse pressure.

The formation of a syrinx in Chiari I malformations has been attributed to the Bernoulli theorem and an increased velocity of CSF flow across the foramen magnum leading to reduced spinal subarachnoid space pressure and subsequent distension of the cord itself. The various theories of generation of a syrinx in spinal cord compression only point to the fact that our understanding of this pathology is still incomplete. This fact is further endorsed by reports of spontaneous regression of syringomyelia even with Chiari malformations. There is therefore a definite association of syringomyelia with cervical spondylosis and reports of clinical and radiological improvement following decompression with or without stabilization points toward an etiological relationship between the two conditions.

Another valid inference from this case is that historically the presence of myelomalacic changes in the cervical cord, namely hyperintensity on T2w images has been associated with poor prognosis following surgery. The prognosis is considered worse if there is a corresponding hypointensity on T1w sequences. We propose that some of these patients with limited local changes may represent a “pre-syrinx” stage, as theorized by Fishbein et al., and may therefore have a better prognosis following treatment.

**Conclusion**

The association of cervical spondylosis with syringomyelia is possibly an etiological one and Cine-MRI with CSF flow studies in addition of evaluation of the CVJ and whole spine to rule out coincidental causes of a syrinx is a valuable diagnostic tool to plan treatment. Decompression of the local pathology often leads to resolution of the syrinx and clinical improvement. The existing concept of cord signal changes in cervical spondylosis being a poor prognostic factor may not be strictly true and a small subset of patients with such changes do show resolution of both clinical symptoms and radiological findings.

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How to cite this article: Bhagavathula Venkata SS, Arimappamagan A, Lafazanos S, Pruthi N. Syringomyelia secondary to cervical spondylosis: Case report and review of literature. J Neurosci Rural Pract 2014;5:78-82.

Source of Support: Nil. Conflict of Interest: None declared.