Syringomyelia associated with cervical spondylosis: A rare condition

Alessandro Landi, Lorenzo Nigro, Nicola Marotta, Cristina Mancarella, Pasquale Donnarumma, Roberto Delfini

Abstract

Spinal spondylosis is an extremely common condition that has only rarely been described as a cause of syringomyelia. We describe a case of syringomyelia associated with cervical spondylosis admitted at our division and treated by our institute. It is the case of a 66-year-old woman. At our observation she was affected by moderate-severe spastic tetraparesis. T2-weighted magnetic resonance imaging (MRI) showed an hyperintense signal within spinal cord from C3 to T1 with a more sharply defined process in the inferior cervical spinal cord. At the same level bulging discs, facets and ligamenta flava hypertrophy determined a compression towards subarachnoid space and spinal cord. Spinal cord compression was more evident in hyperextension rather than flexion. A 4-level laminectomy and subsequent posterior stabilization with intra-articular screws was executed. At 3-mo follow up there was a regression of tetraparesis but motor deficits of the lower limbs residuated. At the same follow up postoperative MRI was executed. It suggested enlargement of the syrinx. Perhaps hyperintensity within spinal cord appeared “bounded” from C3 to C7 with clearer margins. At the level of surgical decompression, subarachnoid space and spinal cord enlargement were also evident.

A review of the literature was executed using PubMed database. The objective of the research was to find an etiopathological theory able to relate syringomyelia with cervical spondylosis. Only 6 articles have been found. At the origin of syringomyelia the mechanisms of compression and instability are proposed. Perhaps other studies assert the importance of subarachnoid space regard cerebrospinal fluid (CSF) dynamic. We postulate that cervical spine instability may be the cause of multiple microtrauma towards spinal cord and consequently may damage spinal cord parenchyma generating myelomalacia and consequently syrinx. Otherwise the hemorrhage within spinal cord central canal can cause an obstruction of CSF outflow, finally generating the syrinx. On the other hand in cervical spondylosis the stenotic elements can affect subarachnoid space. These elements rubbing towards spinal cord during movements of the neck can generate arachnoiditis, subarachnoid hemorrhages and arachnoid adhesions. Analyzing the literature these “complications” of cervical spondylosis are described at the origin of syringomyelia. So surgical decompression, enlarging medullary canal prevents rubbings and contacts between the bone-ligament structures of the spine towards spinal cord and subarachnoid space therefore syringomyelia. Perhaps stabilization is also necessary to prevent instability of the cervical spine at the base of central cord syndrome or syringomyelia. Finally although patients affected by central cord syndrome are usually managed conservatively we advocate, also for them, surgical treatment in cases affected by advanced state of the symptoms and MRI.

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Key words: Syringomyelia; Cervical spondylosis; Syringomyelia surgery; Syringomyelia etiology; Syringomyelia...
Syringomyelia (Gr. Syrinx = tunnel) is a disease characterized by the presence of a cystic tubular cavity within the spinal cord, containing fluid that might be either cerebrospinal fluid (CSF) or indistinguishable from it. It is a very complex disorder with multiple etiologies and a variety of proposed mechanisms of cyst formation. No single theory will cover all instances. Varieties with each of these proposed mechanisms have been described. Syringomyelia is very complex with multiple etiologies and a variety of proposed mechanisms of cyst formation. No single theory will cover all instances. Various with each of these proposed mechanisms have been described. Syringomyelia is considered an exclusion criterion.

Only 6 articles have been found (Table 1). Lucci et al[5] reported 3 cases in their work. In all of them a relation between neurogenic osteoarthropathies of the upper limbs and intramedullary cavitation at spinal computed tomography scan is described but only the third one denotes a relation between “essential” syringomyelia and cervical spondylosis. Lucci admits at the origin of syringomyelia the bony prominence that probably produces ischemia and thus causes the degeneration of the ascending and descending nervous fibers. Kimura et al present the case of a 64-year-old woman. Dynamic magnetic resonance imaging (MRI) revealed instability at C4-C5, spondylosis at C5-C6 and the syrinx extended from C2 to T3 level. It was reduced remarkably after anterior decompression and stabilization. Towards Kimura syringomyelia was caused by intermittent spinal cord compression. Butteriss et al report a case of a 70-year-old man with severe degenerative changes at C5/C6 with a large right paracentral disc-osteophyte complex. An unexpected cord syrinx was noted extending from C6/C7 inferiorly to T6. The patient declined decompressive surgery. Butteriss advocates surgery directed towards relieving the compressive lesion, rather than primary drainage of the syrinx. Kaar et al describe a case of cervical spondylotic myelopathy with instability at C3/C4 and cervicothoracic syrinx at MR imaging. In Kaar’s article the decompression and stabilization, without drainage of the syrinx were considered adequate surgical treatment. Rebai et al describe a case of a 70-year-old patient whose brain and cervical MRI showed syringomyelobulbia with cervical spondylotic myelopathy. Rebai et al proposes a decompressive surgery since extensive cervical laminectomy induced mild clinical improvement and furthermore a second MRI performed showing advanced state of the symptoms and magnetic resonance imaging.

We describe a case of syringomyelia associated with cervical spondylosis admitted at our division and treated by our institute. Moreover, a review of the literature was executed using PubMed database. Objective of the research was to find an etiopathological theory able to relate syringomyelia with cervical spondylosis. We included only case reports about syringomyelia associated with cervical spondylosis. The simultaneous presence of another etiopathological factor at the origin of syringomyelia was considered an exclusion criterion.

### INTRODUCTION

Syringomyelia (Gr. Syrinx = tunnel) is a disease characterized by the presence of a cystic tubular cavity within the spinal cord, containing fluid that might be either cerebrospinal fluid (CSF) or indistinguishable from it. It is a very complex disorder with multiple etiologies and a variety of proposed mechanisms of cyst formation. No single theory will cover all instances. It may develop by various factors. It is most commonly associated with complex hindbrain malformations, such as Chiari malformations, encephalocele and Dandy-Walker cysts. Other causes include postmeningitic and posthemorrhagic hydrocephalus, basilar invagination, spinal arachnoiditis, extraventricular compressions, tethered cord, acquired tonsillar herniation, intramedullary spinal tumours. Acute traumatic cervical spinal stenosis due to fracture or acute severe disc prolapse may result in secondary syrinx formation. Spinal spondylosis is an extremely common condition that has only rarely been described as a cause of syringomyelia. We analyzed the pertinent literature trying to show a possible etiopathogenetic mechanism.

### CASE REPORT

We describe a case of syringomyelia associated with cervical spondylosis admitted at our division and treated by our institute. Moreover, a review of the literature was executed using PubMed database. Objective of the research was to find an etiopathological theory able to relate syringomyelia with cervical spondylosis. We included only case reports about syringomyelia associated with cervical spondylosis. The simultaneous presence of another etiopathological factor at the origin of syringomyelia was considered an exclusion criterion.

### Table 1 Review of the literature

| Author          | Age (yr) | Spondylosis level and sex | Proposed mechanism                                      |
|-----------------|---------|--------------------------|---------------------------------------------------------|
| Kaar et al[4]   | 71, F   | C3-C4                    | Instability of the spine                                 |
| Kimura et al[5] | 64, F   | C4-C5, C6-C7             | Intermittent spinal cord compression                      |
| Rebai et al[6]  | 70, M   | Not specified            | A purely extradural decompression could be sufficient to induce regression of the medullary cavitation. |
| Lucci et al[7]  | 56, M   | C4                       | The bony prominence produces ischemia and thus causes the degeneration of the ascending and descending nervous fibers |
| Butteriss et al[8] | 70, M   | C5-C6                    | Improvement of related symptoms after decompressive surgery |
| Kameyama et al[9] | 59, M   | C3-C4 at C6-C7           | The symptoms of the upper limbs improved after immobilization of the neck |

The table exposes case reports about syringomyelia associated with cervical spondylosis reported in literature. Author, age and sex of the patient, spondylosis level and the proposed mechanism at the base of syringomyelia are mentioned. F: Female; M: Male.

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Landi A, Nigro L, Marotta N, Mancarella C, Donnarumma P, Delfini R. Syringomyelia associated with cervical spondylosis: A rare condition. World J Clin Cases 2013; 1(3): 111-115 Available from: URL: http://www.wjgnet.com/2307-8960/full/v1/i3/111.htm DOI: http://dx.doi.org/10.12998/wjcc.v1.i3.111
The case of a 66-year-old woman, whose presenting symptoms were gait disturbances and loss of balance. These disorders appeared about 9 mo before our observation. At our observation she was affected by moderate-severe spastic tetraparesis. Cervical dynamic-MRI was executed. T2-weighted MRI showed an hyperintense signal within spinal cord from C3 to T1. This process in the central cord resembled “central cord syndrome” with a more sharply defined process in the inferior cervical spinal cord that suggested syrinx (Figure 1). In hyperextension (Figure 1B) bulging discs were evident at the levels C3-C4, C4-C5, C5-C6, C6-C7 and C7-D1. At the same level facets and ligamenta flava hypertrophy determined a compression towards subarachnoid space and spinal cord. Spinal cord compression is more evident in hyperextension rather than flexion. We decided to undergo the patient at the surgical treatment. A 4-level laminectomy and subsequent posterior stabilization with intra-articular screws was executed. In the early post-operative days the patient underwent neuromotor rehabilitation with intra-articular screws was executed. In the early post-operative days the patient underwent neuromotor rehabilitation and slight improvement of tetraparesis was evident. At 3-mo follow up there was regression of tetraparesis but motor deficits of the lower limbs residuated. At the same follow up postoperative MRI was executed. It (Figure 1D) suggested enlargement of the syrinx. Perhaps “central cord syndrome” appeared “bounded” from C3 to C7. At the level of surgical decompression, subarachnoid space and spinal cord enlargement were also evident.

**DISCUSSION**

Already in the 1950’s some authors found a relationship between these two pathological entities. Stern et al. recognized that cervical spondylosis can present sensory and motor symptoms quite similar to the syringomyelic ones. Brain et al. found radiographic lesions of the cervical spine in about 50% of the syringomyelic cases identical to those noted in spondylosis. The possible coexistence of the two diseases was claimed by Smith too. The results of the literature review revealed that compressive mechanism is the major theory at the origin of syringomyelia associated with cervical spondylosis. Al-Mefty et al. proposed that compression causes cystic necrosis (myelomalacia). In a second period as the myelomalacia progresses, the necrotic tissue is phagocytized leaving a secondary cavity (syrinx) within the atrophied spinal cord. Also Uchida et al. conducting a study on a twy/twy mouse, a unique animal that develops spontaneous spinal cord compression without any other reported genetic difference in the anatomy or physiology of the spinal cord, showed that spinal cord mechanical compression is characterized by the loss and exfoliation of anterior horn neurons with progressive spongy degeneration and demyelination in the white matter. Perhaps the extent of demyelination and Wallerian degeneration in the white matter increases proportionately with the magnitude of spinal cord compression. On the other hand, most of the apoptotic cells observed were oligodendrocyte. Though insignificant if compared to acute spinal cord injury, the longitudinally diffuse and extensive pattern of oligodendrocyte apoptosis in twy/twy mouse may be similar to the secondary damage process observed after acute trauma. In Levine’s study at the origin of syringomyelia is described a subarachnoid obstruction. It may result in increasing CSF pressure above the block, compared with below generating a transmural hydrostatic effect with the collapse of vessels within the subarachnoid space above the block, and their dilatation below it. This mechanical stress on the cord parenchyma causes disruption of the blood-brain barrier, which in concert with raised intravascular pressure results in ultrafiltration of crystalloids and accumulation of fluid. Goel state that the fluid may dissect along planes of weakness within the cord resulting in the pathological appearance of a syrinx. It has been postulated that the development...
of high fluid pressure and syrinx formation within the cord may act to counteract the local effect of the primary compressive lesion and as such may be a protective phenomenon. Several authors observing syringomyelia in spinal arachnoiditis, stated that intramedullary cystic degeneration is caused by ischemia due to circulatory disturbance in the subarachnoid space. Also the blockage of CSF pathways around the spinal cord, contribute to formation of cystic cavities. It is often believed that in syringomyelia with spinal arachnoiditis the CSF enters transversally into the syrinx from the blocked subarachnoid space.

Brierley first demonstrated movement of CSF tracers from the subarachnoid space into the spinal cord perivasculary spaces. Then Rennels et al., Wagner et al. and Borson et al. have shown that horseradish peroxidase injected into the subarachnoid space rapidly labels the perivascular spaces of the brain and spinal cord. Rennels et al. proposed that a “paravascular” fluid circulation exists in the nervous system, with an active flow of CSF from the subarachnoid space into the perivasculary spaces around arterioles and continuing through the basal lamina around capillaries. Without direct evidence, they suggested that fluid return to the basal lamina around venules or into the perivasculary space of emerging veins. Milhorat et al. and Cifuentes et al. demonstrated that fluid is capable of moving from the spinal cord interstitial space into the central canal. Milhorat et al. suggested that this mechanism constitutes the “lymphatic” function of the spinal cord. Cho et al. reported that injection of kaolin solution into the spinal subarachnoid space enhanced the extension of intramedullary cavitations in a rabbit model of posttraumatic syringomyelia. Josephson et al. demonstrated spinal cord edema and intramedullary cyst formation after spinal thecal sac constriction in rats. Klekamp et al. produced an interstitial type of edema in the spinal cord by placement of a kaolin-soaked fibrin sponge on the posterior surface of the cat spinal cord at C1 to C2. Kimura et al. reported about a patient affected by pain in the right arm, MRI showed instability at C4-C5 and compression of the spinal cord since a central spinal cord hyperintensity was evident in hypextension. All these characteristics can be found in “central cord syndrome”. He admitted that the longstanding static and dynamic intermittent compression of the spinal cord caused by C4 instability produce disorders of CSF dynamics in the spinal subarachnoid space and associated pooling of an abnormal amount of CSF in the spinal cord parenchyma. Perhaps the sloshing effect of the pulsatile CSF pressure, could make the cavity extend into a rostral and ventral direction. Also Kaar et al. described a case of central cord syndrome since the patient was affected by dissociated sensory loss in the upper left limb, instability at C3-C4 on plains radiographs and T2-weighted MRI showing forward-bulging ligamenta flava and hyperintensity of the central spinal cord in T2-weighted MRI. He attributed the origin of the syrinx to cervical spine instability. Our case presents a more sharply defined process in the inferior cervical spinal cord. We postulate that cervical spine instability may be the cause of multiple microtrauma towards spinal cord and consequently may damage spinal cord parenchyma generating myelomalacia and consequently the syrinx. Another possible theory may supports the hemorrhage within spinal cord central canal as the cause of CSF outflow obstruction, finally generating the syrinx. On the other hand we can assert the importance of subarachnoid space regard CSF dynamic. Several studies show that syringomyelia may be caused by tethered cord, arachnoiditis, arachnoid adhesions or subarachnoid hemorrhages. In cervical spondylosis the stenotic elements can affect this space. Rubbing towards spinal cord during movements of the neck can generate these “complications”. On the other hand cervical spondylosis can generate a block of CSF hydrodynamic within subarachnoid space determining a vascular occlusion of the vessels above the block and consequently a spinal cord ischemic injury. Perhaps cervical spondylosis can directly damage spinal parenchyma like acute spinal trauma. This damage may be proportionally related to the grade of stenosis. So surgical decompression, enlarging medullary canal prevents rubbings and contacts between the bone-ligament structures of the spine towards spinal cord and subarachnoid space.

In conclusion, we can assert that syringomyelia is so rarely associated with cervical spondylosis because there are many compensating mechanisms (arterial, venous, CSF ones), like that of “lymphatic” circulation, so mild and intermittent compression like that found in cervical spondylosis hardly can be associated with syringomyelia unless there are other associated conditions like arachnoid adhesions, post-traumatic arachnoiditis, subarachnoid hemorrhages, Chiari malformations that determine an alteration towards the subarachnoid space. On the other hand, cervical spine instability can generate central cord syndrome or syringomyelia. Although patients affected by central cord syndrome are usually managed conservatively we advocate, also for them, surgical treatment in cases affected by advanced state of the symptoms and MRI.

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Syringomyelia associated with cervical spondylosis

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P-Reviewer Haughton V S-Editor Gou SX L-Editor A E-Editor Ma S