

Spontaneously Resorbed Idiopathic Syringomyelia: A Case Report

Spontan Rezorbe Olan İdiopatik Siringomyeli: Olgu Sunumu

ABSTRACT

A 28-year-old female patient presented with severe neck and back pain in 2003. No abnormality was observed at neurological examination. Syringomyelia in the cervical region was determined at MRI. MRI examination of the cranial, thoracic and lumbosacral regions was normal. The patient refused surgery and was followed up. A control cervical MRI 17 months later showed that the syringomyelia had disappeared. This spontaneously resorbed case of idiopathic syringomyelia, presenting solely due to neck and back pain and with a normal neurological examination, was evaluated in the light of syringomyelia pathogenesis as only three similar cases have been identified in the literature.

KEY WORDS: Idiopathic syringomyelia, Spontaneous resolution, Chiari malformations, Magnetic resonance imaging

ÖZ

Yirmi sekiz yaşında kadın hasta 2003 yılında şiddetli bel ve boyun ağrısı şikâyetiyle başvurdu. Nörolojik muayenesinde herhangi bir anormallik saptanmadı. Hastanın MRI incelemesinde servikal bölgede siringomyeli tespit edildi. Kraniyal, torakal ve lumbosakral bölge MRI incelemeleri normaldi. Hasta cerrahi tedaviyi reddettiği için takibe alındı. On yedi ay sonra yapılan kontrol MRI incelemesinde siringomyelinin kaybolmuş olduğu görüldü. Sadece boyun ve sırt ağrısıyla gelen, nörolojik muayenesi normal olan ve literatür incelemesinde benzer sadece üç olgu olduğu saptanan olgu siringomyeli patogenezi ışığında değerlendirildi.

ANAHTAR SÖZCÜKLER: İdiopatik siringomyeli, Spontan rezolüsyon, Chiari malformasyonu, Manyetik rezonans görüntüleme

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INTRODUCTION

Syringomyelia is a chronic disease characterized by a cavity extending longitudinally inside the spinal cord (4). The pathogenesis of syringomyelia is not fully understood. It may occur after a trauma or secondary to a tumor or spontaneously, but is most frequently seen in Chiari I malformation. The prevalence of syringomyelia in patients with Chiari I malformation is 40%-65% (13, 14). Syringomyelia that develops without trauma, spinal tumor or craniocervical or intracerebral pathology is defined as idiopathic syringomyelia (4). Magnetic resonance imaging (MRI) is the gold standard in the diagnosis and follow-up of syringomyelia (3, 4). The patients may present solely because of neck and back pain or with varying neurological complaints. Such patients are treated conservatively or surgically. The most important criteria in the selection of patients for surgery are the severity of symptoms and a progressive nature (1). There have been a few published cases of idiopathic syringomyelia disappearing spontaneously (4, 16). The spontaneous healing mechanism in idiopathic syringomyelia is still the subject of debate.

CASE REPORT

A 28-year-old woman presented with a complaint of neck and back pain that had begun eight months before. There was no history of trauma that might account for the pain. Neurological examination was normal, and routine laboratory tests were within normal parameters. Cervical lordosis was lost in direct radiograms. Medical treatment with myorelaxants and NSAID's was initiated. MRI was performed as the complaints continued after one month of medical analgesic treatment. A hypointense signal in T1 and a hyperintense signal in T2-weighted sections extending between C5 and T1 and compatible with syringomyelia were encountered (Figure 1). The diagnosis was syringomyelia. Cervical, cerebral, thoracic and lumbosacral MRI scans were also performed and no additional pathology was determined. Surgical treatment was recommended for the severe neck pain resistant to medication but the patient refused surgery. The authors decided to follow-up the patient. Sixteen months later, the patient had severe neck pain for two but the pain resolved completely without treatment. The neurological examination was still normal. Cervical MRI revealed a completely normal spinal cord 16 months after the first admission (Figure 2).



Figure 1: T2-weighted sagittal MR imaging showing syringomyelia extending between C5 and T1.



Figure 2: T2-weighted sagittal MR imaging showing normal spinal cord 17 months after the first MRI.

DISCUSSION

The terms syringohydromyelia, syringomyelia and hydromyelia are used for the definition of fluid cavities in the spinal cord. The term 'hydromyelia' is used to define central canal dilatation. Syringomyelia is typically used in the definition of the central canal dilatation lined with a gliotic wall. In practice, there is no difference between them. A more generic term, syringohydromyelia, or simply syrinx, is used in the definition of enlarged longitudinal fluid cavities inside the spinal cord (2, 3).

In addition to idiopathic causes, syringomyelia is seen together with organic cord diseases (organic syringomyelia) and congenital lesions of foramen magnum or subsequently arising lesions. The symptoms of syringomyelia and/or the syrinx dimension of some patients may resolve without surgical intervention or remain stable (5, 6, 16).

Syrinxes are classified as communicating or non-communicating. In the former, direct communication is between the 4th ventricle and the syrinx. It follows a course together with Chiari I malformation. Non-communicating syrinxes more commonly arise with spinal cord trauma or tumors (2, 3).

The pathogenesis of syringomyelia is still controversial but some hypotheses were proposed. Gardner's hydrodynamic theory (2, 12), William's CSF pressure dissociation theory (13), and the tonsillary piston theory (9) are the most widely accepted hypotheses about the formation of syringomyelia.

The spontaneous resolution of pediatric cases of Chiari I malformation is ascribed to different development speeds of the cranium and central nervous system. As the cranium grows faster than the central nervous system, the posterior fossa structures relax and the foramen magnum is thus eased (14). It is thought that there is restoration of CSF flow in this way and that this leads to the healing of Chiari I malformation and syringomyelia (14).

In the report of two cases, Kazuhiko et al. stated that the MRI showed the cerebellar tonsil descending beneath the foramen magnum, accompanied by C2-T2 syringomyelia, and that MRI six months later demonstrated that the cerebellar tonsils had moved slightly upwards and that the syrinx had disappeared. They attributed the healing to a widening of the CSF distance at the level of the foramen magnum (5).

Klekamp et al. reported a case of an adult with Chiari I malformation and syringomyelia, which resolved spontaneously. They concluded in this report that the spontaneous opening of the CSF pathways in the foramen magnum might be due to the rupture of the arachnoid membranes preventing CSF flow (6).

In the case of a 30-year-old woman with Chiari I malformation and a spontaneously resolving cervicothoracic syrinx, Jack et al. determined at MRI taken 2.5 years later that although the Chiari I malformation had worsened, the cervicothoracic syrinx had disappeared. They attributed the determination at MRI of atrophy in the C4-T2 hemicord to the decompression of the syrinx at this level by desiccation (3). This theory was later supported by MRI in a report by Santoro et al (12). A view supporting this theory was subsequently reported by Milhorat et al. in an analysis of autopsy reports of syringomyelia patients (8).

Hydrocephalus, a likely mechanism of spontaneous healing, is linked to intracranial lesions such as invasive tumors or venous occlusive diseases that affect the flow in the posterior fossa (5, 14). These lesions lead to an increase in intracranial pressure (ICP). The increasing pressure leads in turn to the tonsils being displaced downwards and to syrinx formation. The tonsils resume their former state and there is regulation in the syrinx with the subsequent decrease in ICP (5, 6, 10, 14).

Syringomyelia may develop in cases with small or tight cisterna magna and/or small posterior fossa also known as the Chiari 0 malformation. The tonsils do not change position downwards in this malformation. It is thought that subarachnoid narrowing in the posterior fossa leads to CSF flow disorders. The blockage of CSF causes an increased pressure on the tonsils and this effect plays a role in the development of syringomyelia (5, 7).

Three adult cases with isolated syringomyelia that resolved spontaneously have been reported so far (4, 11, 15). There is only one similar pediatric case in the literature (16). No clear information regarding the development of syringomyelia has been revealed in all these case reports (4, 11, 15, 16).

In their case report, Kastrup et al. suggested that spontaneous resolution might be linked to a rupture that might arise in the spinal cord in connection with increasing pressure during a manoeuvre similar to

the Valsalva manoeuvre. They indicated this hypothesis as the reason for the disappearance of the syrinx at MRI with a worsening of neurological findings. In their reports, however, it is unclear whether there was any finding of spinal cord atrophy on MRI. The syrinx extended from C1 up to the conus in this case (4).

The syrinx was at the C6 level in a case reported by Ozisik et al. It was reported that the syrinx had entirely disappeared in MRI after six years, and there was no abnormal signal in the spinal cord (11).

The syrinx was between C6 and T1 in a case, reported by Vinas et al. They reported that MRI showed a normal spinal cord without any abnormal signal five years later (15).

The syrinx was between C4 and C7 in the 19-month-old girl reported by Yeager et al. There was no abnormality in the posterior fossa and craniocervical region. The syrinx had disappeared and no abnormal signal was observed in the cervical spinal cord in the control MRI two years later (16).

The true mechanism of spontaneous resolution of the idiopathic syrinx cavity in the presented patient is unknown. However, the disappearance of all complaints after two days of severe neck pain and spontaneous resolution of the syrinx suggests that the fistula had developed in the spinal cord and that the syrinx content was drained to the subarachnoid zone. No abnormal signal variation in the cord was observed at MRI.

In conclusion, our case displayed spontaneous resolution like the other cases reported. Our patient was in the adult group and had no history of trauma, Chiari I or Chiari 0 malformation (tight cisterna magna) or intracranial or intraspinal disease. We observed the spontaneous resolution of a completely idiopathic case of syringomyelia with these features. We think that this resolution may be due to a fissure and drainage permitting connection between the syrinx and subarachnoid distance that might exist in the cord.

CONCLUSION

Although it is very unusual, spontaneous resolution of syringomyelia can happen. Conservative treatment should be the first treatment of choice in asymptomatic patients or in patients with moderate complaints.

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