

**Case Report**

***Progressive Neurological Deficit in Adult Idiopathic Syringomyelia (IS):  
Case Report and Literature Review***

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***Abstract***

*A syringomyelia is a clinical entity of any tubular fluid-filled cavity within the spinal cord that causes slowly but relentlessly progressive symptoms as well as expansion of the cavity, which is most commonly associated with Chiari malformation Type I, which due to advancement of imaging techniques has resulted in more incidental idiopathic syringes that are not associated with tumor, trauma, or postinfectious causes. Idiopathic syringomyelia (IS) is a pathological entity in which no overt etiology is evident for a syrinx. In this study, we describe a case in a 45 year-old woman presented with progressive difficulties in walking and also had myelopathic signs evidenced by hyperreflexia in the lower extremities and underwent foramen magnum decompression and C1 laminectomy. Idiopathic syringomyelia is a pathological entity in which no overt etiology is evident for a syrinx. It can be managed successfully by conservative treatment but if there is a progression of neurological deficit, surgical decompression is a mandatory.*

***Keywords:*** *idiopathic syringomyelia, neurological deficit*

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## Defisit Neurologis Progresif pada *Idiopathic Syringomyelia* (IS) Dewasa: Laporan Kasus dan Tinjauan Pustaka

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### Abstrak

*Syringomyelia* adalah kelainan berupa terbentuknya rongga berisi cairan tubular berlebih dalam medulla spinalis yang dapat menyebabkan defisit neurologis progresif, keadaan ini erat hubungannya dengan Chiari malformasi tipe I, dengan kemajuan teknik pencitraan memiliki kemampuan mendeteksi Chiari malformasi yang bukan disebabkan oleh tumor, trauma, atau penyebab *postinfectious*. *Idiopathic Syringomyelia* (IS) adalah keadaan patologis dimana tidak ada etiologi yang jelas untuk terbentuknya syrinx. Pada kasus ini, digambarkan seorang wanita usia 45 tahun, dengan kesulitan defisit neurologis progresif dengan gangguan berjalan disertai *hyperreflexia* di ekstremitas bawah kemudian dilakukan *foramen magnum* dekompresi dan *lamiectomy* C1. IS merupakan suatu keadaan patologis dengan penyebab yang belum diketahui, pada beberapa kasus IS disebabkan karena gangguan aliran LCS (*Liquor Cerebro Spinal*). Penting untuk diketahui pada kasus tertentu IS dapat dilakukan pengelolaan konservatif namun untuk keadaan dengan kelainan neurologis yang progresif dapat dilakukan pembedahan.

**Kata Kunci:** *idiopathic syringomyelia*, defisit neurologis

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### Introduction

Syringomyelia is a chronic disorder involving the spinal cord occurs in some cases without a Chiari malformation or other obvious cause. When the clinical evaluation reveals no evidence of tumor, trauma, or another cause of the syringomyelia, the condition is classified as idiopathic syringomyelia (IS). There are few hypothesis address the pathophysiology of IS. Depending on the source of origin syringomyelia evidence into 3 categories: CSF Entrance from the fourth ventricle, CSF Entrance from the subarachnoid space, extracellular fluid origin and the latest that unifying principles of all types syringomyelia was intramedullary pulse pressure that causes cord distension and subsequent cavitation.<sup>1</sup> Another results showed that IS most frequently associated with Chiari Malformation Type I patients.<sup>2</sup> Some patients with IS with progression symptoms are selected for craniocervical decompression or direct decompression of the syrinx, depending on the clinical findings and the surgical experience and judgment of the surgeon.<sup>1,2,4,5</sup> IS deteriorates progressively if untreated.

### Case

This 33-year-old woman who was otherwise healthy, presented with progressive difficulties in walking. On physical examination that were evidenced by all motoric disturbance (3/5) sensoric disturbance, hyperreflexia in the upper and lower extremities and urinary urgency. MRI T2WI revealed syringomyelia at cervical level with etiology is unclear (Fig1). She underwent craniocervical decompression, which consisted of the following procedures: 1) wide foramen magnum decompression 3 cm x 3 cm via suboccipital craniectomy, 2) peeling of the dura mater, 3) opening of the arachnoid membrane and dissection of arachnoid adhesions over and around the foramen of Magendie and confirmation of sufficient CSF flow and brain pulsation, 4) duraplasty in which we used a lyophilized dura allograft. (Fig 2).<sup>6</sup> This patient did well and showed improvement of upper motoric (5/5) and lower motoric (4/5), urinary urgency and no hypesthesia and hyperreflexia 10 months following surgery.

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**Fig 1. A,B Idiopathic Syringomyelia on T2WI MRI (asterisk)**

**Discussion**

The prevalence of syringomyelia is 8.4 cases per 100,000 population. Approximately 21,000 Americans have syringomyelia, a disorder in which a cyst forms within the spinal cord, resulting in myelopathy. Various theories include the cerebellar piston theory, intramedullary pulse pressure theory, and increased spinal subarachnoid pressure. Idiopathic syringomyelia and Chiari type-I syringomyelia are associated with a small PF with narrow CSF spaces. The etiology of CSF flow disturbances and syringomyelia in idiopathic and Chiari I-type syringomyelia appears to be Posterior Fossa (PF) underdevelopment and neural displacement.

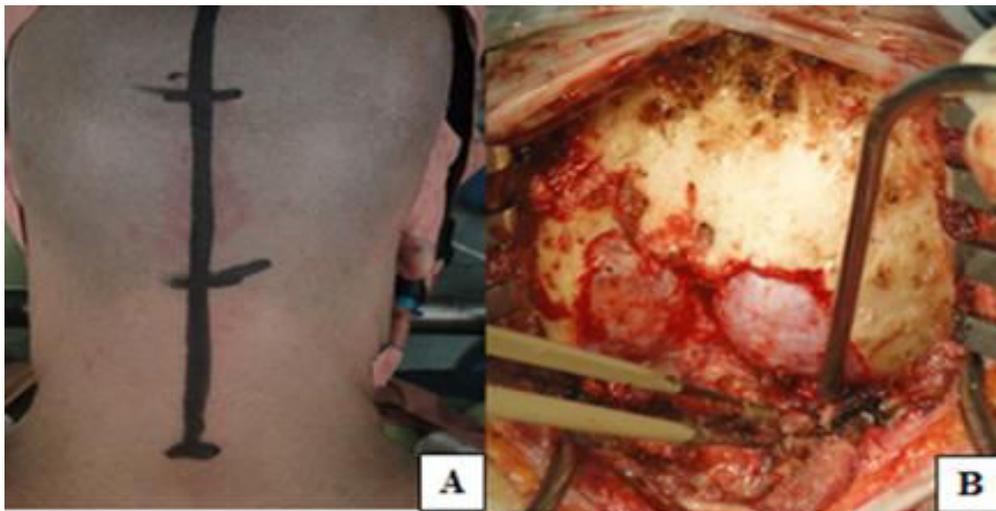
A PF with decreased compliance and narrow CSF spaces promotes the development of accentuated pulsatile CSF subarachnoid pressure waves that promote the development of syringomyelia. Continued progression of symptoms, however, could be approached using decompressive strategies such as C1 laminectomy, lysis of adhesions, and craniocervical decompression, depending on the level of pathology.<sup>1,4,10,11</sup>

The role of CSF flow in the development of IS has not been clarified. Currently, CSF flow obstruction is presumed to cause the accumulation of fluid in the spinal cord. Most research revealed that the syrinx is subject to continuous pulsatile pressure from outside the spinal cord and to sudden rises in pressure associated with Valsalva maneuvers. Cord cavitation causes slow but relentlessly progressive symptoms and signs, depending on its location, extent, and frequent asymmetry.<sup>7,9</sup> The early symptoms usually consist of any combination of motor and sensory dysfunction. Disassociated sensory findings of impaired pain and temperature with the preservation of light touch sensation with “cape-like” suspended sensory loss are the classic

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presentation. Sensory changes usually affect the hands, but a careful examination may disclose a similar deficit affecting the neck, shoulders, upper chest, and back.<sup>7</sup>

This is particularly important because of the good prognosis and excellent chance of recovery if patients are properly managed. The reduced symptoms in patients with IS after cranio-occipital decompression support this presumption.<sup>2</sup>



**Fig 2. A) Linear Skin Incision B) Foramen Magnum Decompression and C1 Laminectomy**

The strategy for treatment patients with IS varies according to the extent of disease progression. Some patients show no signs or symptoms during disease progression for many years and/or the syrinx dimension of some patients may resolve without surgical intervention or remain stable.<sup>8,9</sup> This patient did well and showed improvement of upper motoric (5/5) and lower motoric (4/5), urinary urgency and no hipesthesia and hyperreflexia 10 months following surgery, slight weakness in right lower extremity resulting in possibility of progressive atrophy the lower extremities musculature.

**Conclusion**

Idiopathic syringomyelia is a pathological entity in which no overt etiology is evident for a syrinx. It can be managed successfully by conservative treatment but if there was progression of neurological deficit surgical decompression is a mandatory.

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