ACUTE IDIOPATHIC SYRINGOMYELIA: A CASE REPORT

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There have been isolated reports in the literature of the natural history and less than complete knowledge of the epidemiology and pathophysiology of syringomyelia. This article describes a clinically acute-onset abnormal patient with localized dilatation of the central canal of the spinal cord who had no evidence of present or prior Chiari's malformation, trauma, infection, tumor, or other predisposing pathologic condition. It is suggested that this distinct condition be labeled "acute idiopathic syringomyelia".

Key Words: acute, idiopathic, syringomyelia
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Syringomyelia is a process that involves dilatation of the central canal of the spinal cord or formation of abnormal tubular cavities in its substance. Syringomyelia is not a single disease, but rather a descriptive term for any fluid cavity in the spinal cord. Regardless of the underlying etiology, the symptoms and signs of syringomyelia are related to the location, size, and extent of the cavity. With the advent of magnetic resonance imaging (MRI) and improvements in surgical procedures for decompression of the cavity, syringomyelia is an increasingly recognized cause of disability and even death in patients with clinical symptoms and signs of central spinal cord lesions.

In this article, we describe a case of acute syringomyelia of the cervical cord and term this distinct condition "acute idiopathic syringomyelia" to distinguish it from syringomyelia secondary to causes such as Chiari's malformation, trauma, infection, or neoplasm.

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CASE PRESENTATION

A 19-year-old woman presented with 2-day proximal upper-limb weakness that had developed abruptly and worsened during the first day of her illness. Over the same period, she became aware of unsteadiness of gait. No symptoms of respiratory or gastrointestinal infection were noted. There were no other neurologic symptoms, in particular, no sphincter disturbance or back pain. There was no medical, trauma, or family history of note.

On examination, higher cerebral function and cranial nerves were normal. The strength of upper-limb movements was reduced to a greater extent on the right than on the left (Table 1). Deep tendon reflexes were absent in the arms. In the lower limbs, there was some reduction in strength bilaterally, and bilateral hyperactive deep tendon reflexes with Babinski signs (Table 1). Gait was ataxic, but there was no heel/shin ataxia. Proprioception and discriminatory sensations were normal, but pain and temperature sensations were diminished in the bilateral C5 through T1 dermatomes. Examination of other systems revealed no abnormality. The patient was dependent for activities of daily living, including feeding, grooming, hygiene, dressing, and ambulation (Table 2). Routine hematologic and biochemical investigations, including thyroid function, serum B12, and serologic tests

Table 1. Comparison of motor strength before and after a 1-month comprehensive rehabilitation program (results expressed in muscle strength grade)

	Before management		1 month after management	
	Right	Left	Right	Left
Elbow flexors	2	3	3	5
Wrist extensors	3	3	5	5
Elbow extensors	3	3	3	5
Finger flexors	4	4	5	5
Finger abductors	4	5	5	5
Hip flexors	4	4	5	5
Knee extensors	4	4	5	5
Ankle dorsiflexors	5	5	5	5
Big toe extensors	5	5	5	5
Ankle plantar flexors	5	5	5	5

Table 2. Functional status as evaluated by the Barthel Index before and after a 1-month comprehensive rehabilitation program

	Before management	1 month after management
Feeding	5	10
Grooming	0	5
Bowel control	10	10
Bladder control	10	10
Wheelchair transfer	10	15
Toilet transfer	5	10
Dressing	5	10
Bathing	0	5
Level walking	10	15
Stairs	5	10
Total	60	100

for syphilis, gave normal or negative results. Nerve conduction studies were unremarkable. Cerebrospinal fluid investigation was refused.

In radiologic investigations, cervical spine and chest roentgenography was normal. MRI demonstrated a spinal syrinx between C2 and C6 (Figure 1). The patient subsequently underwent suboccipital craniectomy with C1 laminectomy and C3 through C5 laminectomy to enable craniocervical decompression of the syrinx. Dural vascular engorgement between the C3 and C5 vertebral bodies was found at operation.

Postoperatively, full participation in a comprehensive rehabilitation program was begun, with steady improvements in independence with ambulation at the community level, and independence in activities of daily living. Table 2 depicts the patient's functional status as evaluated by the Barthel index [1]. We compared her functional status before and after the 1-month comprehensive rehabilitation program. Markedly improved motor strength with only a three-fifths strength deficit involving the right proximal upper limb and unchanged sensory deficit were noted. A comparison of motor strength before and after the comprehensive rehabilitation program is shown in Table 1. Twelve months after surgery, the minimal improvement in strength in the right proximal upper limb continued. The sensory deficit remained unchanged. Follow-up MRI revealed a reduction in syrinx size (Figure 2).

DISCUSSION

Syringomyelia has been used to refer to a syndrome caused by cavitation of the spinal cord adjacent to the central canal, or to any cyst of the spinal cord or dilatation of the central

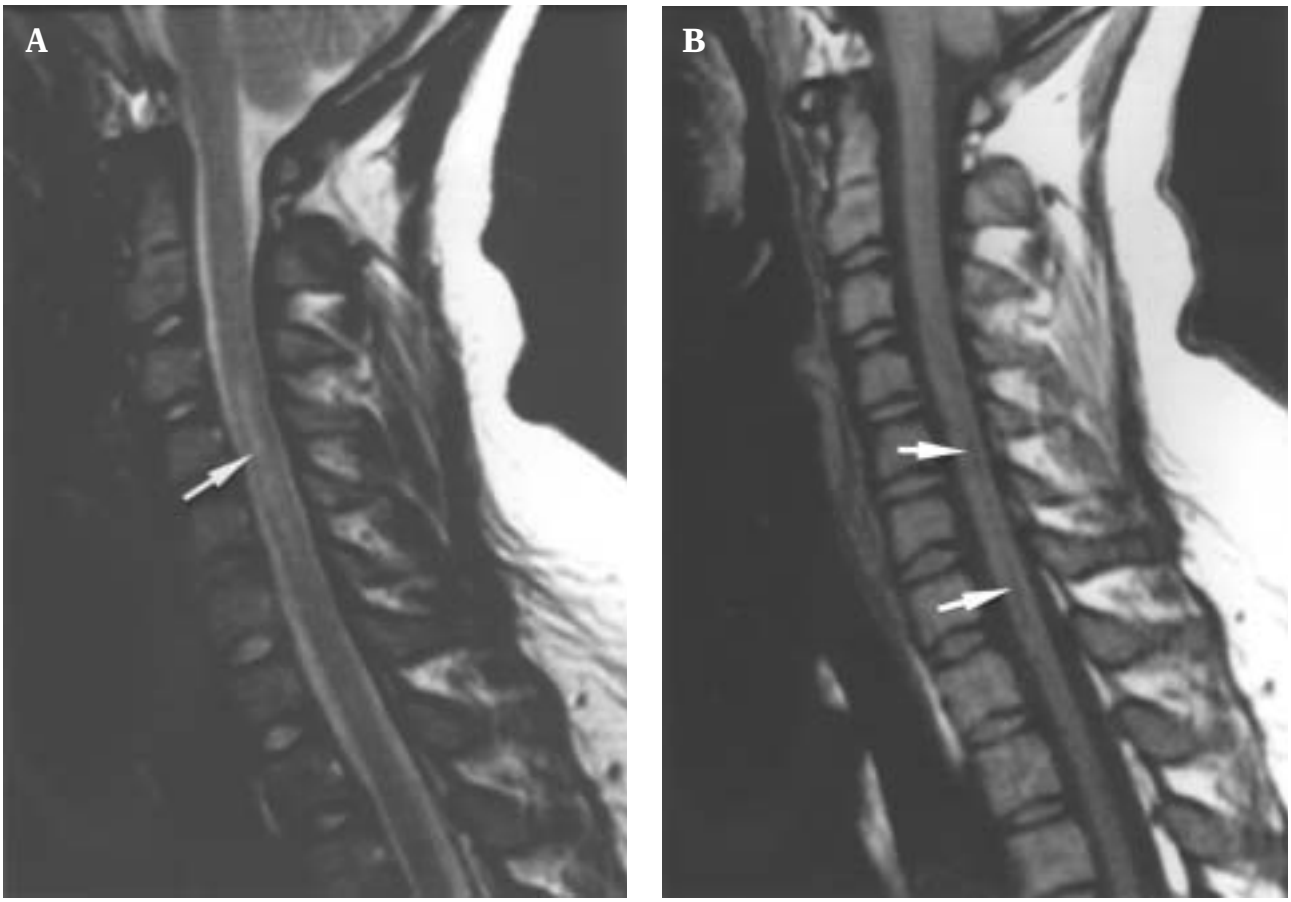

Figure 1. *Preoperative unenhanced sagittal magnetic resonance images of the craniocervical junction and cervical spine show the dilated central canal within the cervical spinal cord (arrows): (A) T2-weighted; (B) T1-weighted.*

canal and/or its extension into the cord substance [2]. The incidence of syringomyelia is unknown, but it was previously thought to be rare [3]. It occurs more frequently in males, and is usually first detected in the third or fourth decades of life. It is uncommon in children or the elderly. Since the advent of computerized tomography (CT) and MRI, there has been an apparent increase in the incidence of syringomyelia. This may represent the detection of previously undiagnosed subclinical cases and not an increased incidence of disease.

The etiology of this disease is variable [4,5]. The dilatation was thought to be primarily developmental in origin, but some postnatal conditions were found to induce the same condition. Barnett, in his seminal monograph on syringomyelia, proposed a more expansive definition and classified the syndrome into five groups according to the presumed causal pathophysiology [6]. The increased intraspinal pressure of such defects in the present patient would result from idiopathic syringomyelia. Unrecognized trauma, coughing, or other such phenomena may initiate development of syringomyelia in the absence of a medical and trauma history. No Chiari's malformation or any skeletal malformation of the craniocervical junction was found.

The pathophysiology of syringomyelia is still debated, and numerous reviews on the subject are available [7,8]. Most articles agree that the syrinx is subject to continuous pulsatile pressure from outside the spinal cord and to sudden rises in pressure associated with coughing, sneezing, and Valsalva maneuvers. Shunting the contents of the syrinx to a low-pressure extraspinal site offers the best chance of maintaining the pressure within the syrinx at a reduced level and dissipating the effects of sudden pressure changes.

There is clinically variable presentation in syringomyelia. Cord cavitation causes slow but relentlessly progressive symptoms and signs, depending on its location, extent, and frequent asymmetry [4]. The symptoms usually consist of any combination of motor and sensory dysfunction. Disassociated sensory findings of impaired pain and

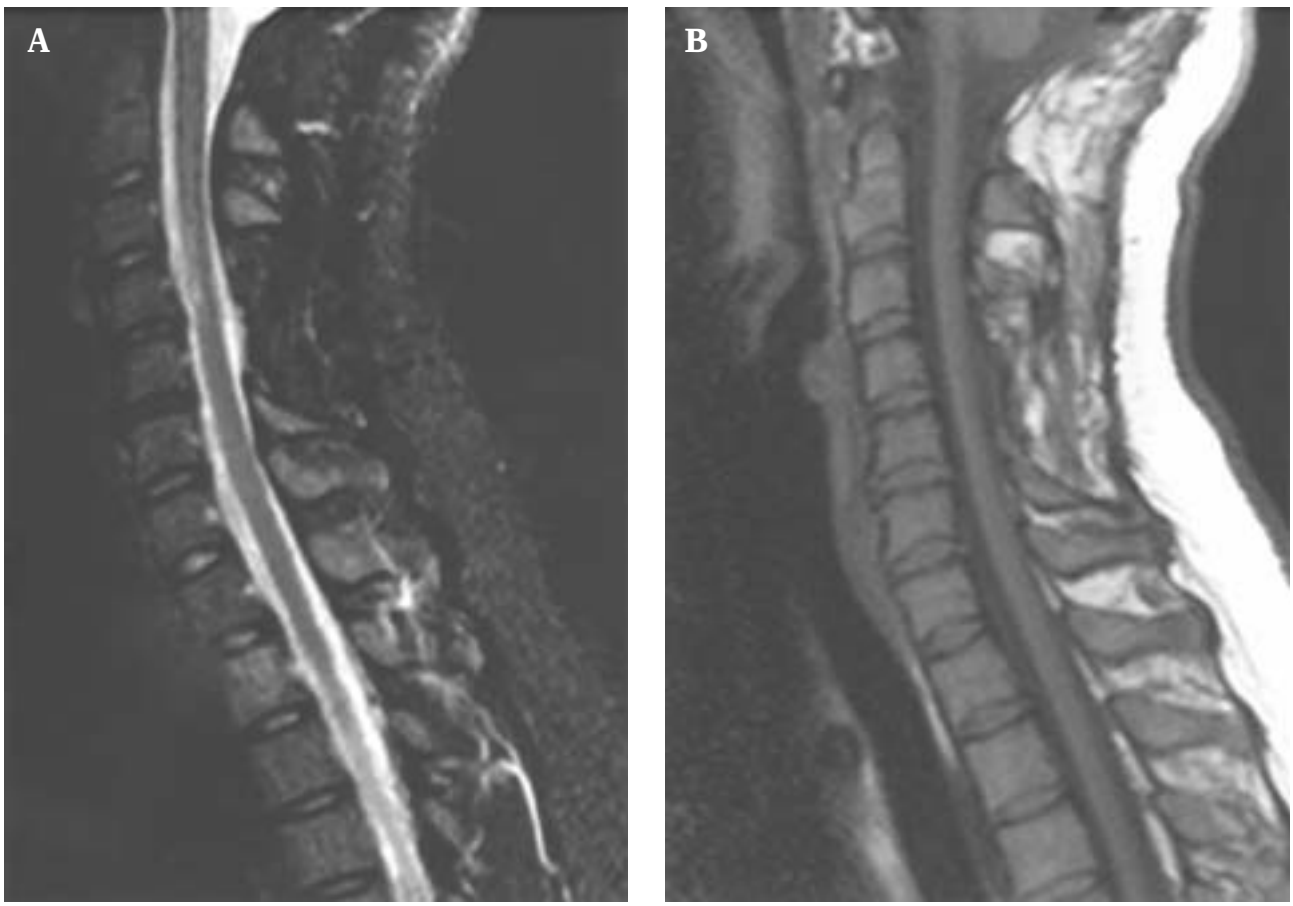

Figure 2. *Postoperative unenhanced sagittal magnetic resonance images of the craniocervical junction and cervical spine (12 months after surgery) demonstrate reduction in syrinx size: (A) T2-weighted; (B) T1-weighted.*

temperature with the preservation of light touch sensation with "cape-like" suspended sensory loss are the classic presentation. The earliest sensory changes usually affect the hands, but a careful examination may disclose a similar deficit affecting the neck, shoulders, upper chest, and back. Motor dysfunction often accompanies the sensory loss, resulting in progressive atrophy of the musculature of the upper extremities. This often manifests in skeletal malformations, principally kyphoscoliosis. The progressive atrophy is seen with other lower motor neuron findings such as muscle fasciculation and areflexia. Later in the disease course, upper motor neuron signs arise in the legs due to involvement of the lateral columns of the spinal cord. The last neurologic functions to be lost are vibratory and position senses in the lower extremities, which occur after the syrinx extends to affect the posterior columns of the spinal cord. Less common manifestations include preganglionic Horner syndrome secondary to involvement of the intermediolateral cell column of the lower cervical and first thoracic segment of the spinal cord containing sympathetic neurons [9–13].

The clinical findings of this case are similar to those reported in previous series. In retrospect, the abrupt onset of proximal upper-limb hypotonic paresis and the cervical dural vascular engorgement found at surgery were the most intriguing features. The clinical presentation was more suggestive of concomitant extrinsic cord compression than of an intramedullary lesion alone. The previous idiopathic syrinx was clinically silent and of indeterminate duration. We believe that syringomyelia developed in our patient as a consequence of localized cord compression due to the distended dural vascular engorgement. It has been postulated that cord compression results in focal necrosis and progressive cavitation [7]. This hypothesis is supported by the patient's sustained clinical improvement following surgical decompression.

Ever since the first case reported by Etienne in 1564, the epidemiology, natural history, pathophysiology, and most prudent form of treatment of syringomyelia have re-

mained controversial. As a clinical entity, idiopathic syringomyelia is a relatively rare condition. To our knowledge, there are no reports describing idiopathic syringomyelia with an acute course. Due to the variation in the natural history, anatomy, surgical treatment, and follow-up periods reported to date, very few firm conclusions can be drawn regarding syringomyelia. Long-duration follow-up is required for a better understanding of all aspects of syringomyelia. We emphasize the need to be aware of this entity. This is particularly important because of the good prognosis and excellent chance of recovery if patients are properly managed.

References

1. Mahoney FI, Barthel D. Functional evaluation: the Barthel index. *Md Med J* 1965;14:61–5.
2. Barnett HJ, Botterell EH, Jousse AT, et al. Progressive myelopathy as a sequel to traumatic paraplegia. *Brain* 1966;89: 159–74.
3. Michals EA, Ramsey RG. Syringomyelia. *Orthop Nurs* 1996;15: 33–40.
4. Williams B. Current concepts of syringomyelia. *Br J Hosp Med* 1970;4:331–42.
5. Sullivan LP, Stears JC, Ringel SP. Etiology of syringomyelia. *Neurology* 1989;39:308.
6. Barnett HJM. The epilogue. In: Barnett HJM, Foster JB, Hudgson P, eds. *Syringomyelia.* London: Saunders, 1973: 302–13.
7. Klekamp J. The pathophysiology of syringomyelia – historical overview and current concept. *Acta Neurochir (Wien)* 2002;144: 649–64.
8. Kyoshima K, Kuroyanagi T, Oya F, et al. Syringomyelia without hindbrain herniation: tight cisterna magna. *J Neurosurg* 2002; 96(2 Suppl):239–49.
9. McIlroy WJ, Richardson JC. Syringomyelia: a clinical review of 75 cases. *Can Med Assoc J* 1965;93:731–4.
10. Cahan LD, Bentson JR. Considerations in the diagnosis and treatment of syringomyelia and the Chiari malformation. *J Neurosurg* 1982;57:24–31.
11. Ellertsson AB. Syringomyelia and other cystic spinal cord lesions. *Acta Neurol Scand* 1969;45:403–17.
12. Faulhauer K, Loew K. The surgical treatment of syringomyelia. Long-term results. *Acta Neurochir (Wien)* 1978;44: 215–22.
13. Van den Bergh R, Hoorens G, Van Calenbergh F. Syringomyelia: a retrospective study. Part I: clinical features. *Acta Neurol Belg* 1990;90:93–9.

急性原發性脊髓空洞症 — 病例報告

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脊髓空洞症為脊髓腔洞擴張以致漸進性之脊髓病變。對於脊髓空洞症的流行病學、病理生理學及自然病程，目前仍未有完整的了解，並只有少量的研究報告。本文報告一無外傷、感染、腫瘤或先天發育缺失病史之急性發病的原發性脊髓空洞症病例。

關鍵詞：急性，原發性，脊髓空洞症

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