

Case Report

Adult syringomyelia associated with Chiari I malformation treated with cervical manipulation: a case report

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Abstract: Syringomyelia associated with Chiari I malformation (SACM) is a rare disease characterized by cavitation of the spinal cord and cerebellar tonsils herniation. Pain, primarily sensory and motor disturbances may present depending on the extent and site of the syrinx and consequent gliosis. Surgery treatments are usually considered as golden standards, while the risk is huge and the results are uncertain. Cervical manipulation has been rarely reported for treating SACM. Here, we report a case of a 35-year-old female patient who was diagnosed with SACM and complained of suboccipital headache and cervicalgia. She was treated for two months, with eight visits using low-amplitude cervical manipulation. At the end of the treatment, the patient's symptoms were relieved obviously, and the imaging showed obvious resolution of syrinxes and spinal cord edema.

Keywords: Syringomyelia, Chiari malformation, pain, manipulation

Introduction

Syringomyelia associated with Chiari I malformation (SACM) is a rare neurological condition characterized by a cystic dilation and expansion of the spinal cord, and cerebellar tonsils' descent of more than 5 mm below the foramen magnum [1]. The mean prevalence of SACM ranges between 2 and 13 per 100,000 inhabitants in different countries, and the ratio of males to females varies between 1:2 and 1:3.7 [2]. SACM can cause typical neurological symptoms and signs, which can be due to the cavitation within the spinal cord, altered cerebrospinal fluid (CSF) flow and compression of central nervous system structures [3]. The quality of life for patients with SACM is generally lower than that of the general population, so this condition continues to present medical and social problems.

Pain is an early and major symptom of SACM, which is usually confined to the head/neck region or upper extremities and can make the patient suffer a lot [4]. Many surgical regimens have been proposed for dealing with this condi-

tion, while lots of them are risky and the results were uncertain. Cervical manipulation has been proved to be a useful intervention in acute cervical pain and radiculopathy, while its use in management of SACM is still rare. In this article, we present a case describing a patient who suffered from SACM and was effectively treated by cervical manipulation.

Case presentation

A 35-year-old female patient was admitted to the Department of Orthopaedic Surgery for the first time, because of 3-month history of suboccipital headache and neck pain. This pain was dull, burning, and occasionally stinging. The onset of pain was gradual, and it could be exacerbated by coughing or sneezing. She rated the intensity of her pain as 8 out of 10 on a numerical rating scale (NRS). Besides, She had no history of tumor, infection, or previous trauma to the spinal cord. On physical examination, upper cervical spine tenderness was found at C2-C5. Cervical range of motion was painful during extension and right rotation. The sense of pain and temperature was decreased in the

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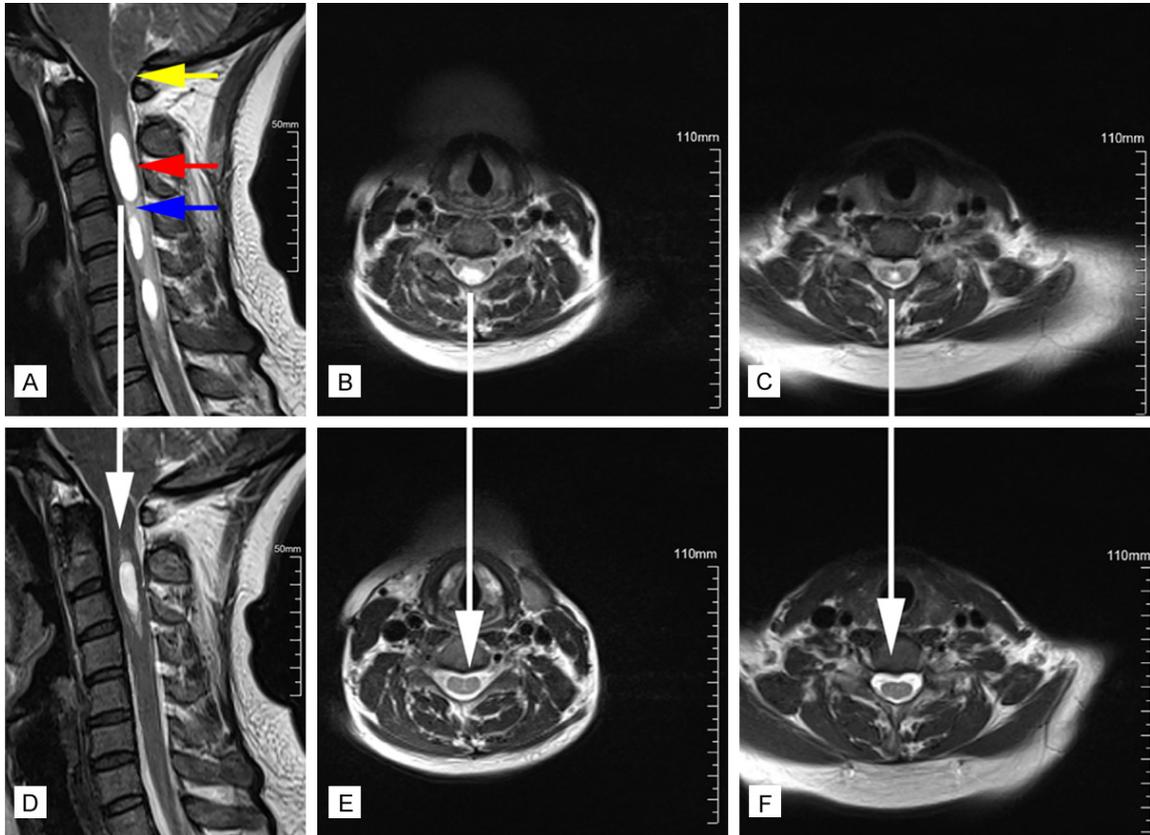


Figure 1. T2-weighted magnetic resonance imaging (MRI) of the patient's cervical spine before (A-C) and after (D-F) treatment. Midsagittal MRI before treatment (A) showed multicystic syringomyelia (red arrow), cerebellar tonsil herniation (yellow arrow), and spinal cord edema (blue arrow), while image after treatment (D) showed resolution of syringomyelia and spinal cord edema (white arrow). Axial MRI at C4 level (B) and C6 level (C) before treatment showed the syrinxes which occupied the spinal cord, while images at C4 level (E) and C6 level (F) after treatment showed collapse of syrinxes (white arrows).

Table 1. Study characteristics and outcomes of reviewed studies

Reference	Year	Study type	Age (y)/Sex	Follow-up	Outcome of manipulation
[14]	2001	Case report	47/female	12 months	Aggravation of symptoms
[15]	2006	Case series	19-21/male	24 months	Aggravation of symptoms
[16]	2014	Case report	34/female	3 months	Relief of symptoms
[17]	2014	Case report	18/female	1 month	Relief of symptoms

radial half of both forearms. Muscle strength was slightly decreased in the right forearm. Upper extremity reflexes were normal bilaterally. Hoffmann's sign was negative on both sides. The magnetic resonance imaging (MRI) showed the multicystic syringomyelia from C3 to C6 (Figure 1A-C), cerebellar tonsil herniation (Figure 1A), and spinal cord edema from C2 to C7 (Figure 1A). Based on above, her diagnosis of SACM was made, and was differentiated from migraine or tension-type headache. At the beginning, we offered some surgery options for

the patient, while considering the risks, she finally gave up. After a consultation with the rehabilitation specialists, she was recommended to take a course of cervical manipulation as a main therapy. She was treated for two months, with eight visits using low-amplitude adjustments (a high-velocity type of joint thrust manipulation) and mobilization (a low-velocity type of joint oscillation) to C2-C6, according to the methods of Bronfort G [5]. The details of the procedure were as follows: First, the patient sat on a chair and rested her elbows on the

desk with the chiropractor standing behind her back. The patient's head was put into a position that included some combination of extension, lateral rotation, lateral flexion and traction. Then, the manipulation took place with a single high-velocity, low amplitude thrust that sent the intervertebral joints slightly beyond the range of normal passive movement. At last, finger-press massage was conducted on bilateral neck muscles, which last for 20 minutes.

At the end of the treatment, the patient's symptoms were relieved obviously and the pain scale decreased to 2/10. The dysesthesia and muscle weakness of forearms were improved too. Radiographically, the MRI reexamination showed good results that the lower two syrinxes collapsed and the size of the top syrinx decreased (**Figure 1D-F**), which we hadn't expected, although the tonsillar descent didn't change apparently (**Figure 1D**). Moreover, it demonstrated the obvious resolution of spinal cord edema (**Figure 1D**). At her 6-month follow-up, there were no recurrences of pains.

Discussion

SACM is a specific condition of central nervous system characterized by syrinx cavity formation in the spinal cord and cerebellar tonsil ectopia. Historically, the term syringomyelia was first applied by Ollivier d'Angers in 1827, and Hans Chiari first described three forms of hindbrain herniation in 1896, which were known as the Chiari malformations [2]. Chiari malformation is estimated to be associated with syringomyelia in as many as 40% of cases [1, 6]. So far, the mechanism that produces SACM has not yet been convincingly explained. The cause of Chiari I malformation may be idiopathic or secondary. Underdevelopment of the posterior fossa and skull base bony structures has been considered as the main cause of primary Chiari I malformation, while secondary causes may include hydrocephalus, congenital vertebral anomaly and arachnoid scarring [7]. In adults, the main cause of syringomyelia is Chiari I malformation, and other causes include trauma, tumor and inflammation. Contemporary theories of syringomyelia pathogenesis have largely focused on the hydrodynamic mechanisms, which can be divided into those that implicate a flow from the fourth ventricle into the central canal and those that involve fluid flowing across the cord parenchyma from the subarachnoid

space, mechanisms limiting outflow, and pressure effects on the cord causing dissection of cord tissue by an existing syrinx [3, 8]. In this case, the patient had no history of tumor, infection, or previous trauma, so primary Chiari malformation was considered. Meanwhile, the coexisting syringomyelia of this patient may be secondary to the Chiari malformation due to the disturbance of free CSF flow at the cranio-cervical junction.

Regardless of sensory or motor deficits, pain is the predominant symptom experienced in 50% to 90% of adult patients with SACM, which may present with occipital or suboccipital headache, cervicalgia or radiculalgia. Hindbrain CSF flow abnormalities has been suggested as the underlying pathomechanism of headaches by the fact that the symptom can be exacerbated by the Valsalva maneuver [9]. Besides, tonsillar ectopia may stimulate nociceptive nerves of posterior fossa and perhaps sensitize medullary or upper cervical pain pathways to produce short-lasting exertional headaches [2, 7]. Moreover, spinothalamic impairment is also necessary for the development of pain in SACM [3]. Generally, surgical treatments of SACM are centered on decompression of the cerebellar tonsils or shunting of the syrinx, which include posterior fossa decompression, suboccipital craniectomy, syringosubarachnoid shunting, and so on [10]. While to what extent will surgery relieve the pain is hard to predict, and every surgery has certain risks or recurrence rate [11]. Some studies report that syrinx that extend to the posterolateral medullary region or an axial medullary occupation greater than 75% is usually associated with poor pain outcome after surgery [9].

Cervical manipulation, as an important chiropractic treatment, has been increasingly used to manage acute and chronic mechanical neck pain or headaches, and was considered to be more effective and safer than medications. In America or Canada, spinal manipulation has become the cornerstone of chiropractic care [12]. While its using in the management of SACM is still controversial [13], as shown in **Table 1**. Leong WK described a patient with asymptomatic SACM who acutely deteriorated after chiropractic manipulation of the cervical spine [14]. Besides, Mäkelä JP reported that the symptomatic SACM in the military conscripts could be aggravated by chiropractic

manipulation [15]. On the contrary, Sergeant AW reported that a patient with headaches and dizziness and a diagnosis of SACM responded positively to chiropractic care [16], and Tieppo Franco V reported that a patient with neck pain and left arm radiculopathy and a diagnosis of SACM was effectively treated with spinal manipulation [17]. There might be some reasons that cervical manipulation worked effectively in this case. First, it might intermittently restore normal CSF circulation at the craniocervical junction through the traction effect, which deflated the syrinx and promoted the resolution of spinal cord edema. Second, it might stimulate mechanoreceptors and reduce the pain pathways. Being a technically demanding procedure, cervical manipulation may lead to such complications as local discomfort, myelopathies or radiculopathies, as well as spinal cord injury [18]. More serious ones are cerebrovascular accidents, such as vertebral artery dissection. Patients with bony abnormalities at occipitocervical junction or previous symptoms of brain stem ischaemia should avoid manipulation [14]. Others have also suggested that malignancy, poorly controlled diabetes mellitus, anticoagulant therapy and infection are absolute contraindications to manipulation [14]. We suggest that cervical manipulation can be considered in patients with symptomatic SACM after the contraindications were excluded. Besides, we recommend that the cervical vascular ultrasound is routinely performed before the manipulation for evaluating the risk of cerebrovascular accidents, and end-range rotation and extension of the cervical spine, which are extremely stressful on the vertebral artery, should be avoided.

Conclusion

Syringomyelia associated with Chiari I malformation (SACM) is a rare condition characterized by cavitation of the spinal cord and cerebellar tonsils herniation. Cervical manipulation can be an effective noninvasive method in the treatment of adult patients with SACM, whose chief complaints were headache and/or neck pain. More cases are needed to further verify the effectiveness of cervical manipulation in treating SACM.

Disclosure of conflict of interest

None.

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