Detection of syringomyelia in a pediatric patient with mild scoliosis: a case report

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Consent: Written consent was obtained from the patient’s mother (as the patient was a minor) to use information and images from her file for this case report.

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It can be challenging to detect syringomyelia in patients with scoliosis, as some cases are mildly symptomatic with little to no neurological deficits. However, a timely diagnosis of syringomyelia is needed to facilitate important treatment considerations. This case report details an 11-year-old female with mild scoliosis and a two-year history of spinal pain that had short-term symptomatic relief from chiropractic treatment. Subtle neurological signs were detected only at re-evaluation, which prompted further investigation with radiographs and subsequent magnetic resonance imaging (MRI). MRI revealed a non-expansile syrinx measuring 3 mm at its widest diameter that extended from C5 to the conus medullaris. The aim of this case is to heighten awareness of the potential diagnostic challenges in patients with syringomyelia and scoliosis. The incidence, pathogenesis, clinical presentation, and management of syringomyelia will be presented to help primary contact providers with appropriate referral and co-management of these patients.

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KEY WORDS: scoliosis, syringomyelia, syrinx, diagnosis, chiropractic, conservative management

La détection de la syringomyélie peut être difficile chez les patients atteints de scoliose, car certains cas sont légèrement symptomatiques, avec peu ou pas de déficits neurologiques. Toutefois, il faut effectuer un diagnostic rapide de la syringomyélie pour faciliter les aspects importants de traitement. Cette étude de cas présente une jeune de 11 ans atteinte de scoliose légère, avec des douleurs vertébrales depuis deux ans qui ont bénéficié d’un soulagement symptomatique à court terme à la suite d’un traitement chiropratique. Des signes neurologiques subtils ont été détectés seulement pendant un nouvel examen, ce qui a incité un examen plus approfondi à l’aide de radiographies et l’imagerie par résonance magnétique (IRM). L’IRM a révélé une syrinx non extensible mesurant au plus 3 mm de diamètre et s’étendant de C5 jusqu’au cône médullaire. Le but de cette étude est d’accroître la sensibilisation aux difficultés potentielles du diagnostic chez les patients atteints de syringomyélie et de scoliose. L’incidence, la pathogenèse, la présentation clinique et la gestion de la syringomyélie seront présentées en vue d’offrir aux fournisseurs des soins primaires des outils pour l’orientation et la cogestion appropriées de ces patients.

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MOTS CLÉS : scoliose, syringomyélie, syrinx, diagnostic, chiropratique, traitement conservateur

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Introduction

Spinal cord anomalies, including syringomyelia, are commonly associated with scoliosis. It has been reported that 25-85% of patients with syringomyelia also have scoliosis. In previous cases, scoliosis was often first classified as idiopathic, and later considered secondary to the syringomyelia when investigated with magnetic resonance imaging (MRI). The pathogenesis related to concomitant syringomyelia and scoliosis remains unclear. However, scoliosis may be a musculoskeletal sign of an underlying syrinx.

The clinical presentation of patients with syringomyelia and scoliosis varies widely. A case series by Emery et al found no neurological symptoms in five patients and mild neurological dysfunction (details not specified) in one subject. Another case series and narrative review found that four patients had reported pain in various regions of the body, while the other four patients had motor weakness, sensory deficits, hyperreflexia and/or urinary urgency. It has been described that the most common presenting symptom is pain, followed by paresthesias, numbness and unnoticed hand injuries, though long tract signs may also occur. In the pediatric population, the location or character of pain does not appear to correlate with the size or location of the syrinx. The variable clinical presentation makes it difficult for health care providers to suspect and diagnose syringomyelia.

Although a diagnostic challenge, it is important to diagnose syringomyelia because there are important considerations for its management. Most cases of syringomyelia remain stable and can respond well with non-operative treatment and monitoring. However, a small proportion of patients with syringomyelia can experience enlargement of the syrinx and progression of neurological symptoms. Previous cases of patients with enlarging syrinxes also had Chiari Malformations and underwent surgery. It is therefore important for health care providers to have an appropriate index of suspicion of underlying spinal cord anomalies for patients with scoliosis.

This case report chronicles the case of an 11-year-old female with scoliosis and chronic spinal pain that had short-term relief with chiropractic care. On re-evaluation by the chiropractor, subtle neurological signs and symptoms prompted additional diagnostic testing. A non-expansile syrinx extending from C5 to the conus medullaris was found on MRI. This case report focuses on the diagnostic challenges of detecting syringomyelia, and highlights potential signs and symptoms of underlying neuroaxis abnormalities associated with scoliosis. The incidence, pathogenesis, clinical presentation, and management of syringomyelia will be discussed to help facilitate the appropriate diagnosis and management of these patients.

Case Report

An 11-year-old female (ethnicity not known) was referred by her family physician to a chiropractic clinic with a two-year history of spinal pain. The spinal pain started insidiously and was located midline from her cervico-thoracic to thoracolumbar junction. The pain was a dull ache that varied in intensity, but appeared worse in the morning and after school. She rated the current intensity as 5-6 out of 10, and reported that the pain had progressively worsened over time. There was no radiation of pain into her lower extremities, but she experienced bilateral leg weakness when running. Aggravating factors included long walks, running, and hot showers, while relieving factors included Tylenol for temporary relief.

The patient also experienced occasional neck pain and bilateral anterior knee pain that appeared unrelated, but was otherwise in good health. Previous lumbar radiographs ordered by the family physician found mild scoliosis at the thoracolumbar junction that was convex to the left in the lumbar region (no measurement given). Her family history included scoliosis and rheumatoid arthritis in her mother. Her history and systems review were unremarkable for any red flags, previous trauma or surgeries.

On observation, there were no signs of deformities, rashes, swelling or warmth. Mild anterior head carriage and bilateral pes planus were noted. Her score on the Beighton scale was 6 out of 9, suggesting flexibility, and there was minimal hyper-extensibility of the skin on the dorsal hand bilaterally. Adam’s test for rib humping was negative. Cervical, thoracic and lumbar motions were full, but active motion produced mild pain in her paraspinal muscles. Active forward flexion at end range in the thoracic and lumbar region reproduced her chief complaint. Hip, knee and ankle ranges of motion were full and pain free bilaterally. Neurological testing of the upper limb, lower limb, abdominal reflexes, and pathological reflexes were unremarkable. Palpation revealed moderate pain...
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Throughout her paraspinal muscles from cervicothoracic junction to thoracolumbar junction, reproducing her chief complaint. Joint motion palpation revealed no joint restrictions, though mild tenderness was present from T1-L2.

The patient was diagnosed with nonspecific back pain and was recommended a trial of chiropractic treatment. A treatment plan was provided over 3-4 weeks with 1-2 visits per week and consisted of education, soft tissue therapy, thoracic and lumbar joint mobilizations, and rehabilitative exercises. Specifically, the treatment included: 1) education on nature of condition, prognosis, reassurance, encouraging mobility and early return to activity; 2) soft tissue therapy to the paraspinal muscles aimed to relieve myofascial tension; 3) low velocity, low amplitude oscillatory mobilizations to the thoracic and lumbar spine; and 4) strengthening the thoracic and lumbar region with exercises including abdominal curl, bird-dog, cat-camel, plank, side plank, and abdominal bracing. The chiropractor also recommended ongoing monitoring for any progression or change in the patient-reported bilateral leg weakness with running.

Over the next month, the patient was treated six times by the chiropractor and experienced mild improvement. On re-evaluation, she reported 30% improvement in pain from treatment, but the pain relief was temporary. The patient still complained of mild thoracic spinal pain and intermittent flare-ups of her low back pain. On examination, lumbar motion was full in all directions, with minimal back pain on active and passive extension. Palpation for joint motion revealed tenderness from T12 to L3 and L5. Spinous percussion was negative for sharp pain or any jump sign, but elicited moderate pain throughout the thoracic and lumbar spine, with the worst pain reported at L2-3 and L5.

Figure 1:
AP lower (A) cervical radiograph revealed scalloping at the left lateral border of the C4 vertebral body (arrow). Lateral radiograph (B) of the cervical spine revealed scalloping of the anterior edges of the spinolaminar at C5, C6 and C7 (arrows), considered to be likely a normal variant.
A repeat neurological examination revealed decreased sensation to crude touch in the left lateral thigh but was bilaterally symmetric in all other dermatomes. Motor strength was 5/5 bilaterally for all lower limb myotomes, though the patient still complained of bilateral leg weakness with running. Deep tendon reflexes were 1+ bilaterally for Patellar reflexes (with the Jendrassik maneuver) and 1+ bilaterally for Achilles reflexes, which were equivalent findings to the first neurological examination. However, after Herron-Pheasant test\textsuperscript{15} was performed, Achilles reflexes became hyperreflexic at 2+ bilaterally, and motor strength had decreased to 4/5 bilaterally in the hip flexors. Based on the neurological findings, the chiropractor ordered a series of full spine radiographs to further assess the scoliosis, examine for any congenital anomalies, and investigate for causes of the patient’s signs and symptoms.

A full spine radiograph revealed scalloping at the left lateral border of the C4 vertebral body (Figure 1A). The anterior edges of the spinolaminar line were also scalloped at C5, C6 and C7 (Figure 1B). The scalloping visualized at the spinolaminar line is likely a normal variant. However, the vertebral body scalloping at C4 warranted a referral for advanced imaging to rule out a space occupying lesion. Postural changes were visualized, including flattened cervical lordosis with a mild anterior shift in the gravitational line, minimal left lateral listing and accentuated thoracic kyphosis. A minimal left thoracolumbar scoliosis was visualized from T10 to T12 that measured 10° by the Cobb method (Figure 2). The lumbar lordosis was mildly accentuated with a Type 1A lumbosacral transitional segment at L5.

A letter documenting the patient’s response to treatment, findings on re-evaluation and radiographic findings was sent to the family physician by the chiropractor. The family physician ordered a full spine MRI to assess for any spinal cord or soft tissue anomalies. Two months later, an MRI revealed that the left lateral vertebral body scalloping at C4 was due to asymmetrical vertebral arteries, with a hypoplastic right artery and compensatory hyperplastic left artery (Figure 3A). The left vertebral artery was at least twice the diameter of the right. There was also a prominence of the central canal indicating an early slightly expansile syrinx from C5 to the level of the conus medullaris. (Figures 3B and 3C) The syrinx measured 3 mm in its maximum dimension in the mid thoracic spine.

The patient was referred by the family physician for a pediatric neurosurgeon consult. A follow-up evaluation and repeat MRI was scheduled by the pediatric neurosurgeon in one year’s time to monitor the syrinx. The patient was advised to continue with physical therapy for symptomatic relief of her back pain. A recommendation was also made for a referral to a pediatric orthopedic surgeon to assess and monitor the scoliosis. A repeat MRI at one
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A year showed the size of her syrinx was unchanged in comparison to her previous scan. Follow-up with the pediatric neurosurgeon a year later revealed no progression in the patient’s symptomatology and a follow up appointment in three years’ time was suggested.

Discussion

Incidence and Etiology:
Syringomyelia is a term that delineates conditions of abnormal fluid cavities within the spinal cord, while syrinx denotes the fluid-filled cavity within the spinal cord parenchyma. Syrinxes can be lined with ependymal or glial cells and are thought to be filled with a derivative of cerebrospinal fluid (CSF). The incidence of syringomyelia in the population was reported to be 8.4 cases per 100,000. The average age of a scoliosis diagnosis was reported to be approximately 8 years and a syrinx diagnosis at 10 years. Idiopathic scoliosis is more prevalent in females, but there appears to be no clear gender predilection for cases of scoliosis and syringomyelia. In addition, previous studies suggest that syringomyelia, scoliosis and Chiari malformations tend to present concurrently. Diagnostic imaging of our patient also revealed a hypoplastic right vertebral artery, a compensatory hyperplastic left vertebral artery, and a transitional lumbosacral segment. To our knowledge, this is the first case report describing these congenital anomalies presenting concomitantly with the syringomyelia. However, it is often believed that congenital anomalies can present in clusters.

The mechanistic relationship between syringomyelia and scoliosis is not well understood. One theory suggests that syringomyelia occurs secondary to the scoliosis. In this theory, it is proposed that there are radicular lesions and CSF imbalance at the convex side of the scoliosis, which give rise to the syrinx. Other authors hypothesize that asymmetric syrinx expansion affects the medial motor nuclei in the anterior horn of the spinal cord. This results in a motor imbalance of the trunk muscles, initiat-

Figure 3:
Axial T2 weighted MRI (A) reveals a hyperplastic left vertebral artery (arrow) with a hypoplastic right vertebral artery. Sagittal T2 weighted MRI (B) of the cervical and thoracic spine demonstrates a syrinx extending from the C5 vertebral body to the conus medullaris (arrow). Axial T2 weighted MRI (C) demonstrates a syrinx within the parenchyma of the spinal cord (arrow).
ing the development of a scoliosis. In comparison to scoliosis and syringomyelia, theories regarding the etiology of post-traumatic syringomyelia are distinctive and have a temporal association between trauma and syringomyelia formation. It is thought that absorption of haematomas, ischemia and oedema secondary to the force during trauma are involved in the pathogenesis of post-traumatic syringomyelia. In our patient, the scoliosis was already present when the syrinx was found on MRI. Therefore, our case report is unable to further elucidate any of these theories related to the development of syringomyelia and scoliosis.

**Assessment and Diagnosis:**
A thorough history and physical examination are important for detecting syringomyelia in patients with scoliosis. The history should focus on inquiring about the wide range of symptoms that may present with syringomyelia. Patients with symptomatic syringomyelia may initially present with pain, and then progress to dissociated loss of pain and temperature with preservation of light touch and proprioception. Patients with syringomyelia and Chiari malformations often present with occipital pain and headaches. Pes cavus, neuropathic joints and back pain may also be present. Neurological signs and symptoms related to syringomyelia can include asymmetric abdominal and deep tendon reflexes, motor atrophy and weakness, spasticity, loss of bladder control, upper motor neuron lesions, sensory changes and wasting of intrinsic muscles of the hand. It is therefore important to also conduct thorough neurological testing during the physical examination.

An appropriate index of suspicion for neuroaxis abnormalities in patients with scoliosis is required during initial assessments and with re-evaluations over time. During the initial assessment, it is important to rule out neurogenic causes prior to classifying a scoliosis as idiopathic. Indications for advanced imaging to rule out neuroaxis abnormalities in patients with scoliosis include atypical curves such as a left thoracic curve, infantile or juvenile onset, pain, neurological deficits, sympathetic disturbances, rapid curve progression and males with a large curve.

However, syringomyelia can present with nonspecific symptoms or without neurological deficits. Some of these patients experience subtle progression of neurological symptoms that may only be detected on re-evaluation. Health care providers need to consider ongoing monitoring, evaluation and advanced imaging when suspecting neuroaxis abnormalities, including syringomyelia.

Our patient had mild signs and symptoms of a syrinx which can often be overlooked, thus making detection difficult. She suffered from two years of back pain before the syrinx was detected. This may have occurred because our patient did not display characteristic signs of a syrinx such as dissociated pain and temperature loss, upper motor neuron lesions, muscle atrophy or neuropathic joints. Arnold-Chiari malformations are frequently found in patients with a syringomyelia and scoliosis, but was absent in our case. The patient also did not present with a left thoracic curve or rapidly progressing curve that have been found to be associated with neuroaxis abnormalities. Contrary to reports in the literature, our patient did not present with pes cavus but rather presented with bilateral pes planus. She did present with unrelenting spinal pain, decreased motor strength and abnormal deep tendon reflexes that guided the chiropractor to suspect an underlying neuroaxis abnormality. The subtle neurological deficits were only detected on re-evaluation by the chiropractor, who had an appropriate index of suspicion of an underlying neuroaxis abnormality.

**Management:**
There are no guidelines to inform the optimal treatment for patients with syringomyelia. A neurosurgical and orthopedic evaluation is warranted for all patients with a syrinx and a scoliosis. A survey of neurosurgeons revealed that most favor surgical intervention when patients present with progression in motor/sensory loss, scoliosis, associated pain and/or size of the syrinx. For patients who are minimally symptomatic or asymptomatic, the majority suggest monitoring with neurological examinations and MRI every 6-12 months. In this case report, our patient had an early non-expandable syrinx prior to the onset of menarche with mild neurological deficits. The neurosurgeon recommended non-operative treatment for our patient, with follow-up imaging at regular intervals to monitor for expansion and curve progression.

Conservative management is considered the first-line treatment for syringomyelia that are not progressive and in absence of surgical indicators. Most minimally symptomatic or asymptomatic syringomyelia remain stable in the short-
term, and monitoring for symptom and curve progression is suggested for these cases. Some syrinxes appear to undergo spontaneous reduction without treatment. In a retrospective review of 27 patients with scoliosis and syringomyelia, syrinxes spontaneously reduced in size by 50% in 14 patients. Most authors suggest that syringomyelia has a slow and benign course, but both rapid neurological progression and spontaneous resolution have also been reported. Ongoing neurological tests and re-evaluation should be included in the management of patients who are being monitored.

It is not clear which conservative interventions should be used in providing symptomatic relief for patients with syrinxes and no neurological symptoms. A narrative review by Roy et al indicated that 10 of 16 cases were treated conservatively, but did not provide the details of the conservative interventions. A review of the chiropractic literature revealed five cases of patients with post-traumatic syringomyelia whose symptoms were treated with chiropractic care. The patients in three of the reports were primarily treated with spinal manipulation at the level of the syrinx with no adverse events. Of these, two case reports included lifestyle recommendations, and one case report included a Chiropractic Biophysics protocol. On the other hand, Busse et al used low force techniques to the spine, stretching, intermittent traction and soft tissue therapy in a patient with post-traumatic syringomyelia. Busse et al cautioned that for cases of post-traumatic syringomyelia, high-velocity, low-amplitude spinal manipulation should be considered an absolute contraindication in the area of the syrinx. It was suggested that there is potential for rupture of the syrinx and damage to the parenchyma of the spinal cord.

In our case, the chiropractor used a trial of joint mobilizations, soft tissue therapy, and exercises to gauge the patient’s response to treatment. This was in light of the patient’s subjective weakness, and progressive nature of the chronic pain, even though the syrinx was not yet detected. The patient reported 30% improvement in spinal pain with four weeks of treatment, but the relief was short-term. There were no adverse events to the conservative treatment reported by our patient. To our knowledge, this is the first case report in the chiropractic literature to describe conservative interventions for relief of spinal pain related to an atraumatic syrinx. Further research is needed to determine the effectiveness of these conservative interventions for the management of syrinxes with minimal neurological deficits.

**Summary:**

Although syringomyelia can be difficult to detect in patients with scoliosis, a timely diagnosis is important to allow for appropriate management. This case report detailed an 11-year-old female with mild scoliosis, and a two-year history of spinal pain. Subtle neurological signs attributed to her syringomyelia were detected only on re-evaluation by the chiropractor. The chiropractor had an appropriate index of clinical suspicion for an underlying neuroaxis abnormality, which prompted radiographs and MRI. Imaging of the patient in this case revealed a non-expansile syrinx spanning from C5 to the conus medullaris. Ongoing monitoring of the minimally progressive syrinx was used to manage this patient. This case report aids in the diagnosis and management of syringomyelia by primary contact providers, including chiropractors.

**References**