

# Cervical Syrinx as a Cause of Shoulder Pain in 2 Athletes

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**Keywords:** syrinx; syringomyelia; shoulder pain; cervical disorder

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The orthopaedic surgeon can be the first specialist to diagnose a neurologic,<sup>13</sup> endocrine,<sup>15,17,19</sup> oncologic,<sup>14</sup> gynecologic,<sup>16</sup> or infectious disease<sup>18</sup> because of the musculoskeletal symptoms associated with nonorthopaedic conditions. In this respect, the orthopaedic clinician must maintain a high index of suspicion for remarkable disease processes outside the orthopaedic differential diagnosis.

Many neurologic conditions are first seen with musculoskeletal symptoms.<sup>13,22,27</sup> The intimate relationship between the neurologic and musculoskeletal systems must be appreciated by the practicing clinician. One such disorder that may have simultaneously presenting neurologic and musculoskeletal symptoms is syringomyelia. Syringomyelia is a condition characterized by the existence of fluid-filled cavities in the spinal cord.<sup>6,29</sup> The syringomyelic cavity, or syrinx, is formed by a disturbance in the normal cerebral spinal fluid (CSF) flow.<sup>6,29</sup>

Although classically associated with pes cavus<sup>29</sup> and scoliosis,<sup>20</sup> a syrinx can have variable presentations. This report describes 2 cases of syrinx presenting with localized symptoms in a single extremity. The consequences of missing the diagnosis of syringomyelia can be devastating because of the possible progression to neuropathic arthropathy.<sup>21,27</sup> Despite its importance and likelihood of being seen by an orthopaedic surgeon, there is no recent review of the diagnosis in orthopaedic literature.

The purpose of this communication is to describe the presentation of a syrinx in an athlete and review the presentation, diagnosis, and treatment of syringomyelia for the practicing orthopaedic clinician. A discussion of the pathophysiology behind syrinx development is complex and well

debated and, therefore, beyond the scope of this article. The interested reader is referred to a recent review by Greitz.<sup>11</sup>

## CASE REPORTS

### Case 1

An 18-year-old, left-hand-dominant, male high school baseball pitcher was seen with arm fatigue and loss of pitch accuracy and velocity. Four months before presentation, he experienced an acute onset of a sharp pain in his left rib cage, which then radiated to his left shoulder and upper extremity while pitching. The patient tolerated the pain for 1 month until he developed changes in pain and temperature sensation in the left upper extremity, highlighted by the patient's description of noting a difference in water temperature from arm to arm while showering. He denied swelling in the extremity, tingling sensations, and nocturnal pain. His medical history included hyperthyroidism.

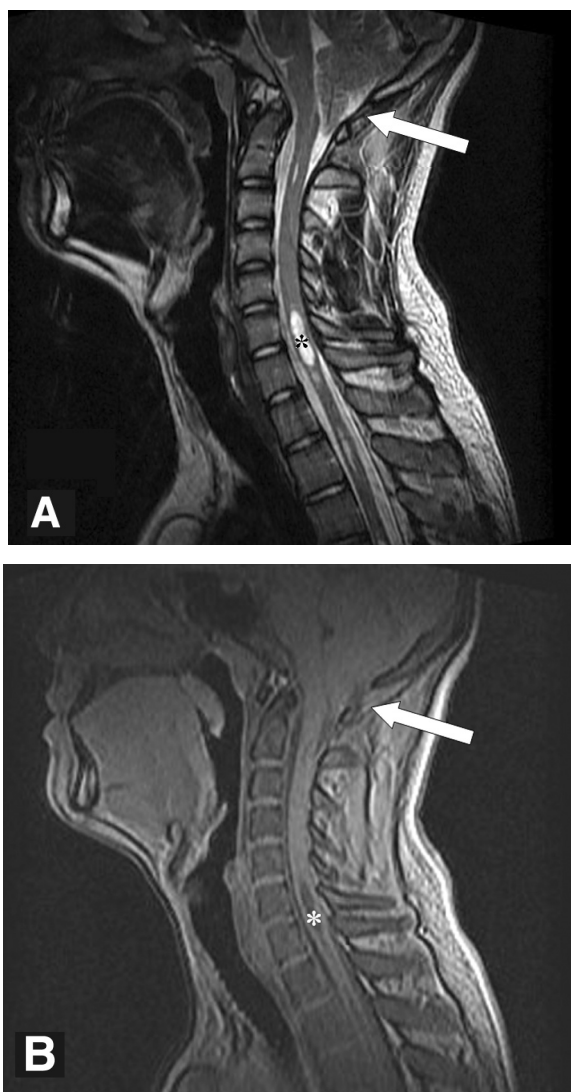
On physical examination, his shoulders were well developed and symmetric, demonstrating full range of motion (ROM) bilaterally with no evidence of scapulothoracic dyskinesia, instability, or labral, acromioclavicular, or rotator cuff abnormalities. There was full ROM of his neck with no tenderness to palpation and a negative Spurling test finding. Manual muscle testing results were normal. Reflexes were brisk, yet symmetric bilaterally, and Hoffman sign results were negative. The examination was notable for only decreased sensation to light touch throughout the entire left upper extremity. Radiograph findings of the neck and shoulder were normal. A cervical spine magnetic resonance imaging (MRI) scan was recommended because of his vague complaints and minor neurologic findings.

The MRI demonstrated a Chiari I malformation with a large cystic fluid collection within the spinal cord consistent with a syrinx (Figure 1). The patient was referred to a neurosurgeon and underwent suboccipital craniotomy and C1 laminectomy with duraplasty. At 11 months postoperatively, he remains completely asymptomatic and has returned to all activities, including pitching college baseball.

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No potential conflict of interest declared.



**Figure 1.** A, T2-weighted magnetic resonance imaging (MRI) scan demonstrating cervical syrinx (asterisk) and tonsillar herniation (arrow). B, T1-weighted MRI scan demonstrating cervical syrinx (asterisk) and tonsillar herniation (arrow).

## Case 2

An otherwise healthy 22-year-old male defensive end college football player was seen with progressive left upper arm pain and weakness after 3 weeks of preseason practice. The pain was described as dull and exacerbated by contact drills. The pain was partially relieved by resting and icing the extremity as well as taking a nonsteroidal anti-inflammatory drug. He additionally complained of weakness with shoulder abduction and external rotation and elbow flexion. The weakness diminished his weight-training abilities and made it impossible for him to hold a 3-point stance with his left arm. He did note mild neck pain but no paresthesias or symptoms distal to the elbow. There were no contralateral arm symptoms with the exception of an inability to distinguish hot versus cold water in his upper extremities.

The patient noted that he experienced this same pain and weakness 18 months before his presentation during spring



**Figure 2.** Index magnetic resonance imaging scan demonstrating the syrinx extending from C5 to T1.

football practice. At that time, he voluntarily withdrew from football, and the symptoms abated until his current presentation. There was no acute event that precipitated his symptoms, and he did not seek care at that time.

On physical examination, he had full neck, shoulder, and elbow ROM. His sensation was intact to light touch, pinprick, vibration, and temperature discrimination throughout both upper extremities. There was very mild atrophy of the left deltoid, supraspinatus, and infraspinatus muscles. Biceps and triceps reflexes were equal bilaterally. Muscle testing revealed a 4 of 5 strength deficit with left shoulder abduction and external rotation, elbow flexion, and finger extension. He had a negative Spurling test finding, and the remaining results of the examination were unremarkable.

Radiographs of his cervical spine revealed a very mild loss of lordosis, and an MRI scan of the cervical and thoracic spine demonstrated minimal cervical spondylosis and no stenosis. There was a syrinx extending from C5 to T1 with a maximal diameter of 3 mm on sagittal sections (Figure 2).

A neurosurgery consultant recommended re-evaluating the syrinx with an MRI at 3 months' follow-up. If the syrinx increased in size or symptoms progressed, surgery would be strongly considered. Otherwise, conservative measures with vigilant observation could be taken. The 3-month follow-up MRI revealed no change in the syrinx, and it was deemed stable. The patient's subjective symptoms completely resolved during the 3-month period, but he had very mild weakness in shoulder abduction and external rotation. There was no biceps or finger extensor weakness.

The patient did not undergo surgical decompression and discontinued his participation in collision sports. He was completely asymptomatic at his latest follow-up of 8 months since presentation.

## DISCUSSION

The presentation of a syrinx can vary. Vague complaints such as pain or altered sensation may be isolated to a single joint

or be found within the whole extremity.<sup>22</sup> Meyer et al<sup>21</sup> demonstrated that the upper extremities are involved in 80% of cases reviewed. The shoulder is the most likely involved joint.<sup>4</sup> Early reviews cited pain as the most common presenting symptom.<sup>28,29</sup> This pain can be localized or generalized to the head, neck, trunk, or limbs and can change in nature or evolve into areas of numbness or tingling over time.<sup>29</sup> More recent reviews have identified swelling of the involved extremity<sup>13</sup> and headache<sup>8</sup> to be just as common as pain in the involved extremity. In a review of 65 adult and 34 pediatric syrinx patients, Ellenbogen et al<sup>8</sup> described this headache to be exacerbated by cough or straining and localized to the nuchal region. Less common presenting symptoms include joint instability, joint stiffness, and neurologic features (changes in sensation or loss of sensation to light touch and proprioception, muscle spasticity, weakness, and atrophy).

The nonspecific presentation and variable signs and symptoms often delay the diagnosis of a syrinx. Moreover, it is a rare condition with an annual estimated incidence of 8.4 per 100 000 people.<sup>6,24</sup> In the most extreme cases, a syrinx is diagnosed only after the development of neuropathic arthropathy.<sup>5,13,23</sup> Neuropathic arthropathy is a serious condition characterized by osseous destruction of the humeral head and glenoid and neurologic deficits.<sup>13</sup> An estimated 25% of syringomyelia cases will progress to neuropathic arthropathy.<sup>21</sup> Hatzis et al<sup>13</sup> reviewed a 28-year clinical experience and identified 7 cases of neuropathic arthropathy. The mean time interval between onset of symptoms and diagnosis was 67 months. They explained that the delay in diagnosis occurred because presentation was often confounded by recent, minor injury. In this review, 6 of the 7 cases were due to syringomyelia.<sup>13</sup>

Frequently, an otherwise asymptomatic syrinx is sometimes discovered after an acute injury. The injury is often minor, ranging from events such as lifting a heavy bag,<sup>22</sup> falling from a chair or bike,<sup>13,29</sup> or reaching to catch an object.<sup>13</sup> The vague symptoms, for example, pain and swelling, and subtle examination findings, such as changes in sensation and proprioception, are often attributed to the acute event, and the syrinx may be found on further diagnostic imaging studies.

Diagnostic tests include plain radiographs of the shoulder and MRI of the cervical spine.<sup>7</sup> The use of MRI provides the resolution for identifying the sharp delineation of a syrinx as well as the evaluation for associated malformations at the craniocervical junction.<sup>9</sup> Should an MRI not be available, computed tomographic myelography can also provide images of sufficient quality to assess for syringomyelia.<sup>12</sup>

The surgical treatment of a syrinx requires the expedient consultation of a neurosurgeon. Conservative management is appropriate if the syringomyelic cavity is stable in size and the patient lacks signs of clinical deterioration or worsening symptoms. Rossier et al<sup>26</sup> demonstrated good results with conservative management in a review of 19 conservatively treated syringomyelia patients. All surgical treatments are aimed at restoring the normal dynamics of CSF flow. Surgical options include expansion or reconstruction of the subarachnoid spaces, bony decompression for kyphosis or scoliosis, and foramen magnum or suboccipital decompression.<sup>1,2,6</sup> Adjunctive procedures such as duraplasty or

arachnoid scar resection have provided improved results.<sup>3</sup> The properly selected surgical treatment provides syrinx resolution and clinical improvement in nearly 100% of cases.<sup>6,10</sup> In the context of sports medicine, it is important to note that after such surgeries, the patient is permitted to return to noncontact, contact, and even collision sports, with the exception of wrestling in which there may be direct contact with the suboccipital area.

A review of the literature outside of neurosurgical publications provided 2 published case reports on syringomyelia in the context of orthopaedic sports medicine.<sup>25,27</sup> The objective of this report is to highlight the subtleties of the clinical presentation of symptoms in these 2 patients and the importance of maintaining an appropriate index of suspicion for other underlying pathologic disorders. While symptoms of syrinx are not classic for subtle instability, there were some similarities to symptoms often seen in the throwing athlete. If the patients had not mentioned temperature asymmetries of the upper extremities, one could have easily misdiagnosed the athlete's symptoms. As most patients present late with neuropathic shoulder findings, it is important for the sports medicine specialist to be cognizant of this condition.

## CONCLUSION

The orthopaedic surgeon must maintain proper vigilance for many nonorthopaedic conditions, particularly those of neurologic origin. Syringomyelia can have a vague presentation that often includes subtle physical examination findings which may overlap other musculoskeletal complaints. The diagnosis may be overlooked because these physical examination findings are often attributed to a recent minor injury or event. Diagnosis is best confirmed through MRI. Treatment of a syrinx requires the consultation of a neurosurgeon. Surgical intervention focuses on re-establishing normal flow of the CSF and is very successful in resolving the syrinx and decreasing symptoms.

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