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Idiopathic Thoracic Intradural Spinal Arachnoid Cyst Associated with Syringomyelia: a Case Report and Literature Review

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We report an adult woman who presented with lower limb weakness and dysesthesia caused by a thoracic spinal arachnoid cyst associated with syringomyelia. Before the diagnosis was made by thoracic magnetic resonance imaging, she was diagnosed with lumbar spondylosis and underwent lumbar spinal surgery; however, her symptoms were not relieved. The patient received multilevel laminectomy and cyst resection after thoracic pathology was identified. An upper motor neuron lesion, although rare, should be considered a differential diagnosis when leg symptoms and signs develop. For patients with leg weakness and pain, we emphasize a detailed neurological examination to localize the lesion to a specific spinal region. (Tw J Phys Med Rehabil 2018; 46(2): 107 - 112)

Key Words: Arachnoid cyst, syringomyelia, spine, drop foot

INTRODUCTION

Spinal arachnoid cysts are rare and can cause neurological deficits through spinal cord compression. Some result in secondary syringomyelia, which can also produce neurological symptoms. Most spinal arachnoid cysts are located in the thoracic spine.[1] Clinically, thoracic spinal canal lesions can be difficult to diagnose because they have similar clinical features to lumbar spine lesions.[2]

We report a patient with drop foot and leg dysesthesia who was initially diagnosed with lumbar radiculopathies and underwent surgery for lumbar retrolisthesis. However, the illness was later correctly diagnosed as an intradural thoracic arachnoid cyst with syringomyelia. Here we review the literature on arachnoid cysts, syringomyelia, and how to differentiate between upper and lower motor neuron lesions when leg symptoms are present.

CASE REPORT

A 57-year-old woman presented with left drop foot in 2012, describing a “heavy and tight” sensation in her left big toe. These symptoms worsened, and she
developed the same sensory symptoms in the right sole. In 2014, at a local community hospital, plain radiography of the lumbar vertebrae revealed retrolisthesis at L4-L5 and lumbar spine magnetic resonance imaging (MRI) showed spinal canal stenosis from L4-S1. Hence, she underwent L4-S1 decompression laminectomy and fusion on September 2, 2014. However, her symptoms continued to worsen after surgery, with sensory symptoms extending to the left upper back and right knee. A thoracic spine MRI on November 1, 2014, revealed a long segmental cystic lesion of cerebrospinal fluid (CSF) signal intensity at intradural space at the T7-T9 level, suggesting an arachnoid cyst with diffuse cord compression and spinal cord atrophy at the T8-T9 level (Figure 1). The spinal cord was displaced to the right posterior side, and syringomyelia at T9-T12 level was also seen (Figure 1).

Due to the persistent symptoms and gait instability, the patient underwent T8-L1 total laminectomy, cyst resection, and T10/11/12/L1 internal fixation at a medical center on May 25, 2016. The histopathology reported as an arachnoid cyst. The postoperative course was uneventful, and the patient experienced a relief of pain symptoms.

The patient was admitted to our hospital for rehabilitation on September 22, 2016. At admission, a neurologic examination showed grade 2-4/5 lower-extremity strength, decreased knee and ankle reflexes, and negative Babinski’s sign bilaterally. Sensory examination revealed hypoesthesia to pain and light touch below the seventh thoracic dermatome. Proprioception was intact and there was no sphincter disturbance. Nerve conduction studies (NCS) and electromyography (EMG) were performed on October 7, 2016, and the results were compatible with polyradiculopathies, including bilateral L2-S1 roots, with active axonal denervation. She started rehabilitation, including land-based and water-based therapy. She was able to stand without help and walk a few meters with assistance at discharge.

Figure 1. A, T2-weighted magnetic resonance imaging (MRI) scan of the thoracic spine, sagittal section demonstrating an arachnoid cyst (arrow), and a syrinx (curved arrow) caudal to the cyst. B, T1-weighted MRI with contrast, and C, T2-weighted MRI axial section at T8-9 level, showing the intramural feature.
DISCUSSION

While spinal arachnoid cysts are relatively uncommon, the actual incidence is unknown. Most are idiopathic, and the rest are secondary to inflammation, in which the causes could be meningitis, trauma, postoperative complications, or hemorrhage.\(^\text{1-3}\) Spinal arachnoid cysts are classified into three types: Type I, extradural cysts without nerve root involvement; Type II, extradural cysts with nerve root fibers; and Type III, intradural cysts.\(^\text{4}\)

The mechanism of syringomyelia formation is considered to be the disturbance of CSF flow, and could be related to Chiari malformation, spinal hemorrhage, or infection.\(^\text{1,2}\) Compared with extradural cysts, syringomyelia is more commonly associated with intradural cysts.\(^\text{5}\) The incidence of syringomyelia resulting from a spinal arachnoid cyst was previously regarded to be rare.\(^\text{1}\) Tucer et al. (2013) reviewed patients with both an idiopathic spinal arachnoid cyst and syringomyelia, with a total of 32 reported cases.\(^\text{6}\) However, there are reports contradicting these data. A case series in 2003 reported seven patients who presented with arachnoid cysts to have syrinx out of the 21 examined patients,\(^\text{5}\) while another case series showed only one patient to have an idiopathic arachnoid cyst to have syrinx in 10 patients.\(^\text{7}\) Moreover, another large case series published in 2018 revealed that two in the 14 examined patients presented with syrinx.\(^\text{8}\) The highest incidence is seen in the report from Klekamp et al., in which 50 of the 109 studied patients with an idiopathic arachnoid cyst had syrinx, while 18 of 21 patients with a secondary arachnoid cyst had syrinx. The rate of intradural arachnoid cysts with accompanying syringomyelia is much higher than in other reports, which might be due to referral bias.\(^\text{2}\) The actual incidence of syrinx in patients with arachnoid cyst is still unclear.

Most arachnoid cysts are located at the thoracic spine (71-100%) and are in the posterior subarachnoid space (60-100%). Most syrinxes form caudal to the arachnoid cyst.\(^\text{1,5,7,9}\) In patients with syringomyelia and idiopathic arachnoid cysts, the most common presenting symptoms are weakness, gait difficulty, back pain, radicular pain, sensation disturbance, bladder, and bowel dysfunction.\(^\text{1-3,6,7,10}\) These symptoms could be attributed to compression of the spinal cord or nerve roots by arachnoid cysts or syrinx. Wang et al. (2003) reported that ventral intradural cysts are likely to cause weakness and myelopathic symptoms, and dorsally located cysts more commonly present with numbness and neuropathic pain.\(^\text{5}\) Our case presents with lower extremity weakness, sensory disturbance, and gait difficulty, consistent with thoracic spinal lesions. The best method of diagnosis of arachnoid cysts is MRI, which has greater diagnostic value than computed tomography myelography, which is helpful to prove the presence of communication of the cyst with the subarachnoid space.\(^\text{2}\)

Treatment for arachnoid cyst include resection, fenestration of the cyst into the CSF spaces, marsupialization, or cyst drainage with shunting.\(^\text{2,10}\) Some authors consider cyst resection as the best choice, while others believe the optimal treatment is still not identified because of the rare entity of the disease.\(^\text{8,10}\) Some suggest complete cyst resection for cysts extending no more than three spinal segments and partial resection for cysts extending over four or more segments.\(^\text{2}\) During the operation, fluoroscopy could help localization.\(^\text{8}\) Intraoperative sonography could be used to detect the size and morphology of the cyst and could prevent excess neural exposure.\(^\text{1,5,8}\) Somatosensory and motor evoked potentials could be utilized for prevention of nerve injury.\(^\text{8}\)

Resection of the primary arachnoid cysts often results in favorable outcomes for both motor and sensory symptoms, despite the presence of an associated syrinx. The formation of syrinx is attributed to the arachnoid cyst, which causes alterations in CSF flow within the spinal subarachnoid space; therefore, the syrinx would regress spontaneously after the arachnoid cyst subsides.\(^\text{2,8}\) However, as for the postoperative outcome of secondary arachnoid cysts, only pain showed moderate improvement. Therefore, surgery should only be recommended if neurological progression or severe spinal cord compression exists. Duroplasty has been a generally accepted principle to prevent spinal cord tethering and CSF obstruction, and patients who undergo duraplasty are more likely to experience pain relief.\(^\text{2,5,8}\)

As for ventral arachnoid cysts, as present in our case, they are more difficult to remove than those located
dorsally. Since aggressive cyst removal could cause spinal cord retraction, an alternative option is cyst shunting or fenestration, and syrinx shunting could also be considered.\textsuperscript{[2,5]} With review of the literature, syringomyelia could be managed by shunting, arachnolysis, decompression (i.e., laminectomy and duroplasty), and myelotomy.\textsuperscript{[11-14]} However, optimal treatment remains controversial. It is generally accepted that if there is a primary source of the syringomyelia, the primary lesion should be managed first.\textsuperscript{[11-14]}

Our patient had onset of drop foot and leg dysesthesia, and was ascribed to lumbar radiculopathies before thoracic myelopathy was identified. As the NCS/EMG examination performed 4 years from her first symptoms revealed many active denervation signs in muscles innervated by lumbar-sacral roots, the denervation could result from the caudally progressing syrinx, which invaded lumbar roots. The symptoms she developed later might have been a result of both upper and lower motor neuron lesions.

The most common cause of drop foot is peripheral neuropathy, followed by other lower motor neuron disorders; central causes, including brain or spinal cord lesion, are relatively uncommon, but should not be ignored. Besides the brain, the whole spinal cord should be assessed when considering upper motor neuron disease.\textsuperscript{[15,16]} Thoracic myelopathy can masquerade as lumbar radiculopathies, and leg pain or weakness can be the first symptom of thoracic cord compression.\textsuperscript{[17-20]} Some patients have undergone an unnecessary lumbar operation before the thoracic cord compression was diagnosed.\textsuperscript{[21]}

To give a correct diagnosis for a patient with leg weakness and dysesthesia, it is necessary to be careful of whether any myelopathic or long-tract sign exist, including provocative signs and hyperreflexia of deep tendon reflex. Provocative signs include Hoffman sign, inverted brachioradialis reflex (finger flexor contraction with a diminished brachioradialis reflex after elicitation), Babinski sign, and sustained clonus.\textsuperscript{[22-25]} Alternatively, we should consider that myelopathic signs could be masked by co-existing lower motor neuron disorders, such as diabetic neuropathy and lumbar radiculopathy, and the rate of being masked was 21% for cervical myelopathy.\textsuperscript{[22]} Another study showed that 85% of patients with cervical myelopathy presented with hyperreflexia, and only 44% had Babinski sign.\textsuperscript{[23]} Although more invasive, epidural block could be used to localize the lesion that causes symptoms, given that co-existing lower motor disorder is not uncommon.\textsuperscript{[24]}

Considering that every myelopathic sign is important and could be masked by lower motor disorders, both sensory level examination and pain characteristics are necessary. The pain induced by myelopathy is called funicular pain or tract pain, which has some different features from root pain. Funicular pain tends to be continuous, diffuse, symmetrical, and poorly localized, instead of localizing as dermatome. The subjective description of tract pain is typically deep, hot, cold, burning, boring, aching with superimposed sharp sensations, but not sharp or stabbing. The pain could be contralateral to cord compression.\textsuperscript{[24]} Besides, the pain cannot be exacerbated by stretching the root, like the straight leg raising test.\textsuperscript{[21,24]}

For our patient, the presentation of pain and weakness in her legs resulted in multiple lumbar imaging studies. She did not receive thoracic spine imaging until 3 years following her initial symptoms. The present case stresses that for patients with leg weakness and pain, a detailed neurological examination should be performed to localize the lesion to a specific spinal region. This could help prioritize diagnostic imaging decisions as well as facilitate treatment and minimize irreversible neurological damages.

**CONCLUSION**

Thoracic myelopathy is relatively uncommon and can mimic other diseases, such as lumbar spinal lesions and peripheral neuropathy. There is a need for detailed neurological examination for complaints of leg weakness or disturbed sensation, to be able to provide proper care for patients with this disorder.

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Spinal arachnoid cyst with syringomyelia


原發性脊髓蛛網膜囊腫及併發之脊髓空洞症－病例報告及文獻回顧

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本文描述一名女性病患因胸椎脊髓蛛網膜囊腫及其伴隨之脊髓空洞症而導致下肢無力及感覺異常。此病患起初症狀為小腿感覺異常及垂足，經影像診斷為腰椎滑脫，故行腰椎椎板切除及融合術。術後症狀仍未改善且異常感覺延伸至上背部，才由胸椎磁振造影發現此蛛網膜囊腫及脊髓空洞症，並經病理切片確診。之後此病人行胸腰椎多節椎板切除併囊腫切除，術後持續復健。

雖然小腿的症狀較少由上運動神經元病灶引起，但仍需列入鑑別診斷中。臨床上面對因腿部症狀而求診的病患，為找到真正的病灶，必須執行詳細的神經學檢查，方能開立更精確的影像學檢查、以及時解決病患的問題並避免不必要的手術。（台灣復健醫誌 2018；46(2)：107-112）

關鍵詞：脊髓蛛網膜囊腫(Arachnoid cyst)、脊髓空洞症(syringomyelia)、脊椎(spine)、垂足(drop foot)