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Case Report

Unilateral Knee Pain as Clinical Presentation of Spinal Dural Arteriovenous Fistula：A Case Report

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Spinal dural arteriovenous fistula (SDAVF) is a rare disease. The principal clinical manifestation of SDAVF is myelopathy, with variable but nonspecific symptoms including progressive weakness of the lower extremities, back pain, bowel and bladder dysfunction, and impotence. Diagnosis is often delayed. Knee pain as the chief complaint of SDAVF is rare. The present case report describes a 49-year-old man with a history of SDAVF, for which he had twice received endovascular embolization, who presented with profound pain in the right knee when attending the study rehabilitation clinic. He had no history of trauma. Physical examination revealed upper motor neuron signs in bilateral lower extremities. Imaging studies of the right knee, including X-ray and sonography, revealed no structural lesion to which his clinical presentation could be attributed. Nerve conduction velocity and electromyography examination showed no evidence of radiculopathy or peripheral neuropathy. Digital subtraction angiography (DSA) identified recurrent SDAVF. It was, thus, believed that the patient's profound right knee pain was the presentation of persistent myelopathy caused by recurrent SDAVF. Early diagnosis and treatment of SDAVF can improve patient's prognosis. Treatment options include endovascular embolization and open spinal surgery. When treating patients with SDAVF, physicians should keep in mind that although two-thirds of these patients can experience motor recovery, only one-third show improvements in sensory disturbances, and pain might persist. (Tw J Phys Med Rehabil 2012; 40(2): 85 - 90)

Key Words: spinal dural arteriovenous fistula, myelopathy, spinal cord vascular malformation, knee pain

INTRODUCTION

Spinal cord vascular malformations can be classified into dural arteriovenous fistulas, arteriovenous malformations, hemangioblastoma, cavernous malformation, and spinal aneurysms.[1] Among these 5 types, spinal dural arteriovenous fistula (SDAVF) is the most common, constituting approximately 60% to 80% of spinal vascular malformations. However, it remains rare and underdiagnosed.[2] These malformations are acquired and consist of a single arterial feeder that develops a fistula to the spinal venous circulation, resulting in perimedullary venous hypertension, subsequently impaired spinal cord venous drainage, and hypoperfusion of the spinal cord.[3,4] The shunt usually locates within the dorsal surface near the dural root sleeve in the thoracolumbar region.[4] Men in their fifth or sixth decades are the age group most commonly affected by SDAVF. The initial clinical presentation is nonspecific, including gait disturbance, progressive

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lower extremity weakness, and sensory disturbance (pain, paresthesia, tightness, and symmetrical or asymmetrical sensory loss). The clinical symptoms can be aggravated by exercise, long-term standing, bending forward, or Valsalva maneuver.[4,5] Bowel and bladder incontinence, sexual dysfunction, or paraplegia can accompany the worsening myelopathy. Diagnosis is often delayed until the development of micturition problems or paraplegia, with a mean interval of 15 months between onset of symptoms and diagnosis.[6] The present case report describes a man with a history of SDAVF, for which he had twice received endovascular embolization, who presented with unilateral knee pain as the clinical presentation of recurrent SDAVF.

CASE REPORT

A 49-year-old man working as a police officer visited the neurosurgical clinic with the chief complaint of progressive weakness and tightness sensation in both lower legs, which he had experienced for approximately 5 months. He also presented with intermittent claudication which was exacerbated after long-term walking or bending forward, and relieved after rest. Mild urinary incontinence developed gradually, within 2 months to 3 months of the onset of bilateral lower limb weakness. Magnetic resonance imaging (MRI) showed SDAVF (Figure 1), which was confirmed by other imaging modalities including computed tomography angiography, magnetic resonance angiography (MRA), and digital subtraction angiography (DSA). Digital subtraction angiography identified the fistula from the right T8 radicular artery branching from the right intercostal artery (Figure 2). The patient underwent transarterial embolization (TAE) thereafter. Gait disturbance, muscle weakness, and incontinence improved after TAE, but the tightness sensation in the lower limbs persisted. Bilateral knee pain developed gradually after the first TAE. The plain film of the bilateral knee was examined at neurosurgical and orthopedic clinics, but physicians were unable to identify the cause of the pain. Seven months after the first TAE, DSA was arranged for further evaluation because of worsening muscle spasms and weakness of bilateral lower limbs. This revealed recurrent SDAVF from the right T9 radicular artery branching from the right T9 intercostal artery (Figure 3). The patient, therefore, underwent a second TAE. However, his clinical symptoms did not show any significant improvement after this procedure. Two months after the second TAE, the patient revisited the clinic because of worsening and excruciating right knee pain.

Physical examination by palpation revealed a normal range of motion in the right knee and no anatomic abnormalities. In manual muscle tests, lower limb strength was 4/5 in the knee extensor bilaterally and 5/5 in all other muscle groups. Deep tendon reflexes (DTR) were 3+ in the left lower extremity and 4+ in the right lower extremity. Clonus was elicited in the right lower extremity and plantar reflexes were equivocal bilaterally. Because of the patient's elusive and intractable right knee pain, imaging studies including X-ray and sonography of the right knee were performed but provided normal findings. Nerve conduction velocity and electromyography (NCV/EMG) examination provided no evidence of radiculopathy or peripheral neuropathy. Myelopathy caused by residual or recurrent SDAVF was, thus, suspected. Digital subtraction angiography was repeated and demonstrated recurrent SDAVF from bilateral T7 radicular arteries (Figure 4); MRA revealed decreased cord edema, compared with the previous images. After discussion with the neurosurgeon, conservative treatment was preferred because of the small size of the recurrent fistula. Thus, antispastic muscle relaxants were prescribed along with physical therapies, including stretch exercises and hot pack to treat hypertonicity of the right lower extremity. However, these treatments displayed limited effectiveness. Figure 5 shows the complete disease course of this patient which is presented in a flow chart.

DISCUSSION

Unilateral knee pain is a common complaint at the rehabilitation clinic, with the patient's history and physical examination results guiding the differential diagnosis. The most common underlying entities are knee joint degenerative diseases associated with soft tissue or other musculoskeletal injury. In the present case, X-ray and soft tissue sonography did not detect any abnormalities. The patient underwent further NCV/EMG examination to rule out radiculopathy, which is another cause of knee pain. The examination showed no evidence of radiculopathy or peripheral neuropathy. The possibility of recurrence of underlying SDAVF was, thus, considered.

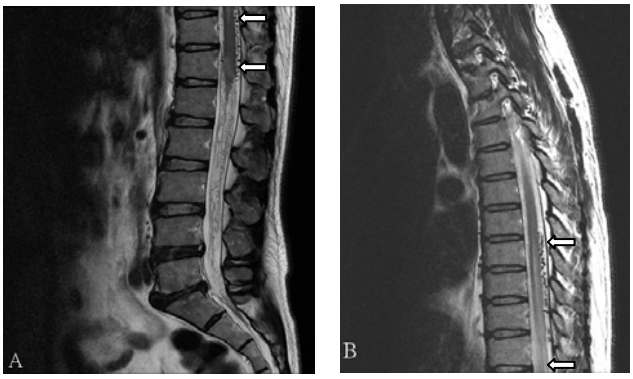

Figure 1. Sagittal T2-weighted MRI at (A) thoracolumbar and (B) thoracic level showed cord swelling and cord edema with serpentine and dilated perimedullary veins (white arrows).

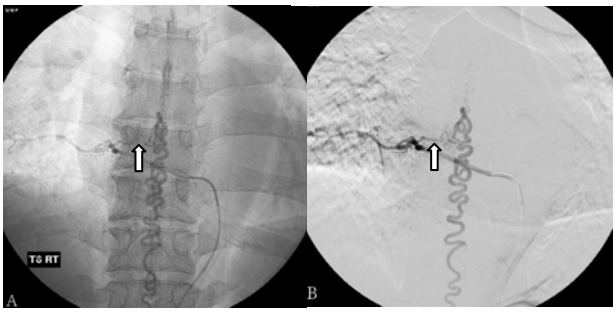

Figure 2. (A、B) Spinal angiography after injection of the right T8 intercostal artery revealed an arteriovenous fistula with shunting to engorged perimedullary veins (white arrows).

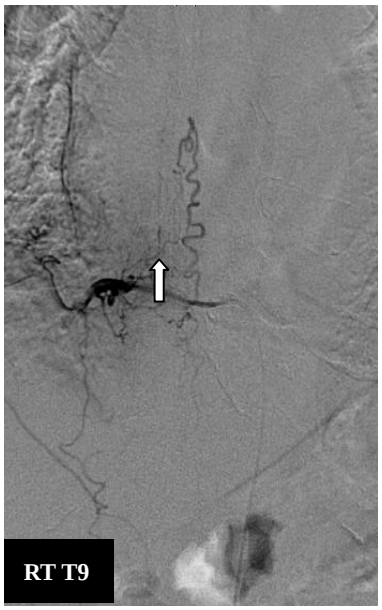

Figure 3. Spinal angiography after injection of the right T9 intercostal artery demonstrates a recurrent arteriovenous fistula with engorged perimedullary veins (white arrow).

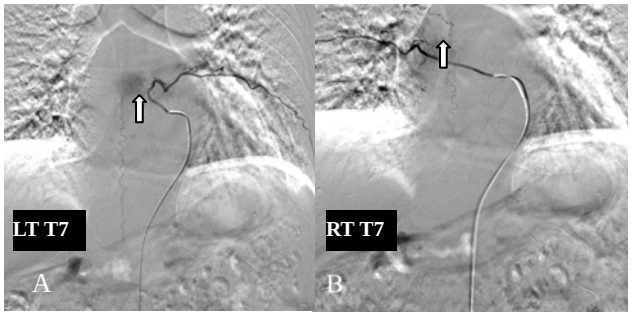

Figure 4. (A、B) Spinal angiography after injection of the bilateral T7 intercostal arteries demonstrates small recurrent arteriovenous fistula (white arrows).

According to the reviewed literature, in recent years, neurologists, neurosurgeons, and neuroimaging specialists have gained significant understanding of SDAVF. However, it remains unfamiliar to and frequently ignored by other physicians. Because of its rarity and nonspecific clinical symptoms, diagnosis of SDAVF is often delayed. Misdiagnosis as polyradiculopathy, polyneuropathy, transverse myelitis, degenerative disc disease, acute or chronic inflammatory demyelinating polyneuropathy or spinal tumor can also occur.[2,4,7-9] As the myelopathy worsens, sphincter disturbance might develop in combination with upper motor neuron signs. This is indicative that the location of the lesion is not in the roots or nerves.[6] McKeon et al reported 3 cases of SDAVF initially misdiagnosed as longitudinally extensive transverse myelitis. Paraplegia worsened after corticosteroid treatment.[5] The outcome of SDAVF is dependent on pretreatment disabilities and the duration between onset of symptoms and diagnosis; therefore, early diagnosis and treatment is imperative. When treated appropriately, SDAVF is curable.[4,7-9]

Imaging modalities play a crucial role in the diagnosis of SDAVF and posttreatment follow-up. Magnetic resonance imaging is helpful for the identification of cord edema, and dilated and serpentine vessels on the cord surface.[9] Magnetic resonance angiography can localize the lesion and provide detailed information on the size and course of the fistula. This can provide guidance to the following spinal angiography, which is the gold standard for SDAVF diagnosis.[2] In the present case, follow-up MRI after TAE revealed decreased cord edema in comparison with previous images. Instead of demonstrating

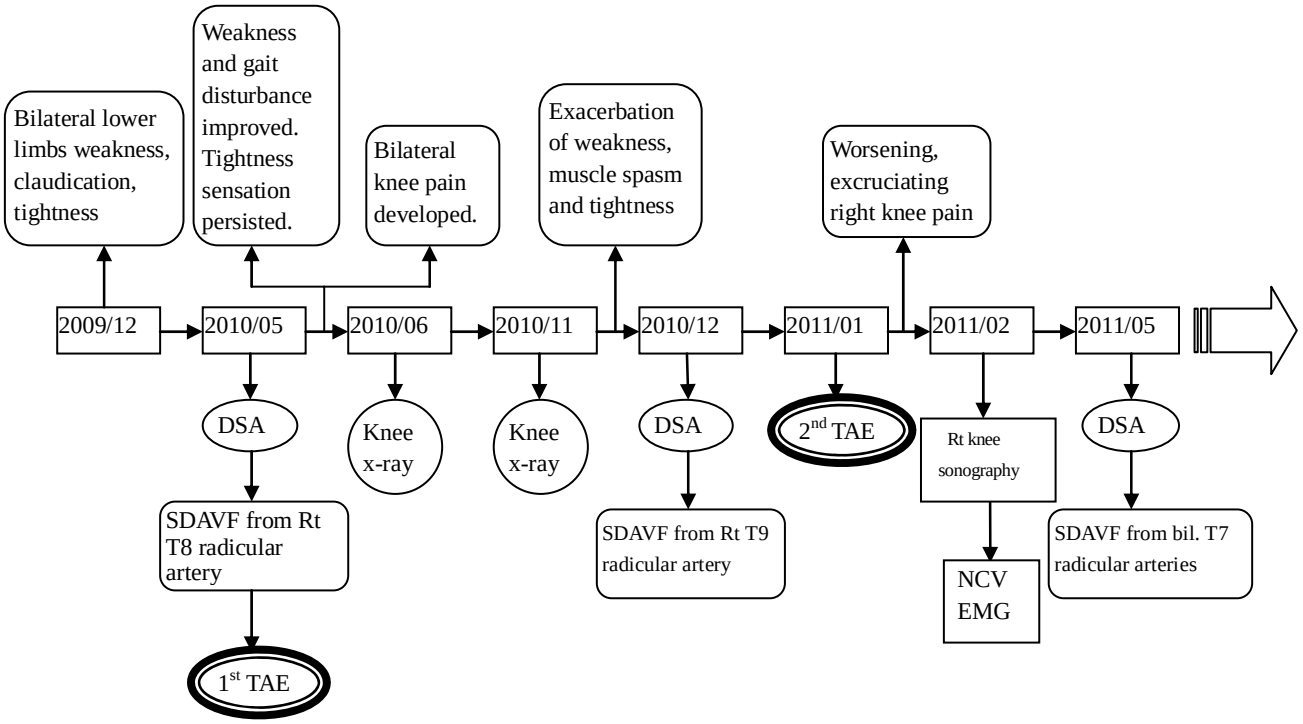

Figure 5. The flow chart of entire course of our case. (DSA: digital subtraction angiography; TAE: transarterial embolization; SDAVF: Spinal dural arteriovenous fistula; NCV: Nerve conduction velocity; EMG: electromyography)

concurrent improvement, the patient's clinical symptoms deteriorated, with increasing pain and in the right adjacent muscle tightness knee. Some correlation between MRI and clinical outcome exists; however, previous studies have mainly reported that MRI changes after treatment have no relation to clinical outcome.[7,10,11]

Among the common clinical symptoms of SDAVF, gait disturbance and muscle strength respond more favorably to treatment than pain, micturition difficulties, and muscle spasms. Erectile dysfunction and sphincter disturbances are seldom reversible, and pain might persist and become refractory to treatment.[4,8,9,12] The same pattern manifested in the present case: his pain and muscle spasms failed to recover and worsened after treatment. Jellema et al described a "release phenomenon" as a possible explanation for this trend, occurring by reperfusion to the permanently damaged area of spinal cord which leads to subsequently unfavorable changes in connections between the remaining nerve cells.[12]

Treatment options for SDAVF include endovascular embolization and open spinal surgery. Endovascular embolization is less invasive than surgery and can be performed during spinal angiography. However, in previous studies, the rate of success of endovascular embolization has varied, whereas surgery obliteration resulted in almost complete occlusion of the fistula.[9] The reported success rate for endovascular embolization has varied between 25% and 90%. The reported success rate for complete occlusion of the fistula following surgical intervention, however, is 98%.[2,9,13] Because of recent improvement in techniques for the endovascular procedure and its low invasiveness, endovascular embolization remains the first treatment option in the clinical setting. Surgical intervention is indicated when an endovascular approach is unfeasible, or if the arterial feeder of the fistula also supplies the anterior or posterior spinal artery, making spinal cord ischemia a possible complication from embolization.[2,4] If recanalization of the previously embolized fistula develops at follow-up, surgery should be considered for effective resolution.[2,13] However, the surgical indications for SDAVF have yet to be well-established. In the present case, clinical symptoms of muscle weakness and gait disturbance improved significantly after the first TAE, despite the persistence of the tightness sensation. Because of the previously successful treatment experience and the safety and feasibility of TAE,

the neurosurgeon selected TAE instead of open spinal surgery as the treatment option following the diagnosis of recurrence of SDAVF from the right T9 radicular artery. Conservative treatment was preferred for the patient's knee pain arising from recurrence of SDAVF from bilateral T7 radicular arteries because of the small size of the recurrent fistula.

CONCLUSION

Unilateral knee pain is a common presentation in the rehabilitation setting. However, as a major complaint of residual or recurrent SDAVF, it is extremely rare. Spinal dural arteriovenous fistula is a treatable cause of progressive myelopathy. Its early diagnosis and treatment is, therefore, critical. The initial presentation of SDAVF is nonspecific and it might mimic other neuromuscular disease. When a patient presents with knee pain associated with gait disturbance, sensory disturbance, and sphincter or sexual dysfunction, the physician should always consider SDAVF as one of the differential diagnoses.

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脊髓硬膜動靜脈瘻管以單側膝關節疼痛爲臨床表現：病例報告

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脊髓硬膜動靜脈瘻管(spinal dural arteriovenous fistula)是很罕見的疾病，臨床表現多爲脊髓病變(myelopathy)，症狀多樣化且不具特異性，包含漸進性下肢無力，背痛，大小便功能異常或陽痿。初期常會延遲診斷，而以膝關節疼痛爲主訴的案例更是少見。我們報告一名 49 歲，具有脊髓硬膜動靜脈瘻管病史並接受過兩次血管栓塞術治療的男性患者，因急性右側膝關節疼痛僵硬至復健科求診。病史詢問後發現無外傷病史，理學檢查發現雙側下肢出現上運動神經元現象(upper motor neuron sign)。膝關節相關影像學檢查含 X 光及軟組織超音波均無可解釋臨床症狀的結構性病變，下肢肌電神經檢查之結果無神經根或周邊神經病變，血管攝影顯示硬膜動靜脈瘻管復發，因此推論該病患右膝關節疼痛應爲瘻管復發所造成脊髓病變之表現。早期診斷及早期治療有較好的預後，治療可以選擇血管栓塞術或脊椎手術。治療後約有三分之二的病人運動功能可恢復，但只有約三分之一的病人感覺異常會改善，此外疼痛的情形則可能一直持續，值得臨床醫師注意。（台灣復健醫誌 2012；40(2)：85 - 90）

關鍵詞：脊髓硬膜動靜脈瘻管(spinal dural arteriovenous fistula)，脊髓病變(myelopathy)，脊髓血管病變(spinal cord vascular malformation)，膝痛(knee pain)

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