12세 환아에서 교대마비 증상으로 나타난 경막외 혈종을 동반한 경부 척수 경막 동정맥루 1례

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Lower Cervical Spinal Dural Arteriovenous Fistula with Extradural Hematoma in a 12-year-old Boy Presenting with Alternating Hemiplegia

The majority of spinal dural arteriovenous fistulas (SDAVFs) develop in elderly patients and are located between the lower region of the thoracic spine and the upper region of the lumbar spine. Herein, we report the case of a 12-year-old boy diagnosed with lower cervical SDAVF and a concurrent extradural hematoma. The patient complained of weakness in his upper and lower extremities with alternating hemiplegia as an initial symptom. A spinal magnetic resonance imaging (MRI) scan revealed an extradural mass lesion with a heterogeneous signal intensity at the C7-T2 level. The mass had low signal intensity at the T2 level. The timely use of an MRI scan facilitated a prompt diagnosis. The patient's symptoms drastically improved following a decompression laminectomy. The pathologic findings of the specimen were consistent with arteriovenous fistula. We report this case because it is clinically important due to the rarity of lower cervical SDAVF in pediatric patients. Furthermore, early diagnosis and treatment are required to prevent irreversible neurological deficit.

Key Words: Arteriovenous Fistula, Vascular Malformations, Hemiplegia, Hematoma, Epidural, Cranial

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Introduction

Although spinal dural arteriovenous fistula (SDAVF) is rare, it is the most frequent vascular malformation of the spine. SDAVF predominantly affects men older than 50 years of age. The most frequent locations are the lower region of the thoracic spine and the upper region of the lumbar spine. Thus, low cervical lesions are extremely rare. The exact etiology of SDAVF is unknown; however, it is considered an acquired vascular malformation. The diagnosis of SDAVF is difficult due to the rarity of the disease and the misleading nature of the initial symptoms.

In this report, we describe the case of a 12-year-old boy with lower cervical SDAVF. The patient complained of weakness in his upper and lower extremities with alternating hemiplegia as an initial symptom and was diagnosed with SDAVF.

We announced this report as a poster presentation in the Annual Symposium of the Korean Child Neurology Society, 2014.

Submitted: 31 October, 2015

Revised: 2 November, 2015

Accepted: 22 December, 2015

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Case Report

A 12-year-old boy presented to the pediatric department with a progressive 5-day history of left-arm and right-leg weakness. Five days before this visit, he felt a tingling sensation in his fourth and fifth left fingers and cramps in his left hand and both legs. He also experienced chest discomfort. Symptoms were aggravated upon anterior flexion of the neck. Chest radiograph revealed no pathology or unexpected abnormalities. Echocardiographic measurements were within standard normal limits. Two days after initial presentation, weakness developed in his left arm and right leg. It steadily progressed, resulting in left-leg weakness. There were no symptoms of cranial nerve injury.

This patient was the third eldest sibling and born at 41 weeks and 4 days of gestation via normal spontaneous vaginal delivery at a local hospital. His birth weight was 3,160 g. Neither he nor any of his family members had a past history of medical or neurological disease.

On physical and neurological examination, Medical Research Council (MRC) grade 2 and MRC grade 4, measure of motor function, were noted in the distal and the proximal part of the right lower extremity, respectively. Strength of the left lower extremity changed from MRC grade 5 to MRC grade 4. MRC grades 4 and 5 were noted in the left and right upper extremities, respectively. Deep tendon reflexes were normoactive or mildly elevated in the upper extremities and elevated in the lower extremities. Sustained clonus of both ankle joints were noted. Babinski's sign was present on both sides. Sensory tests showed normal results.

His laboratory investigations on admission were unremarkable.

A spinal magnetic resonance imaging (MRI) scan revealed an extradural hematoma with a heterogeneous signal intensity lesion at the level of C7-T2 and low signal intensity at the level of T2. The hypointense signal was related to epidural venous ectasia. Additionally, the spinal MRI scan suggested the possibility of an underlying vascular malformation with venous thrombosis (Fig. 1).

The patient had open surgery. Partial rupture of the ligament was seen at the level of C6 and C7. Any signs of muscle or other connective tissue contusion were not noted. A hematoma, probably due to vascular malformation, was located between T1 and T2. This patient had C7-T1 laminotomy and T2 decompression laminectomy. After the decompression laminectomy and excision of hematoma, his symptoms drastically improved. The pathologic findings showed vascular malformation features including an artery with remarkable smooth muscle hyperplasia, collagen deposition, reduplication of the internal lamina, and a vein with an arterialized and thickened collagen coated wall. The pathologic

diagnosis was arteriovenous fistula (Fig. 2).

The interval between the first symptom and diagnosis by the spinal MRI scan was 5 days. The patient underwent laminectomy on the day of diagnosis. The postoperative course was uneventful. His condition improved and he regained normal function of extremities.

Discussion

In 1926, Foix and Alajouanine¹⁾ first described SDAVF as "myélite nécrotique subaiguë" (subacute necrotizing myelitis), which was discovered during the autopsy of two young patients with ascending myelopathy. Spetzler et al.²⁾ proposed a classification system for spinal cord vascular lesions that utilized three primary categories: neoplasms, aneurysms, and arteriovenous lesions. Arteriovenous lesions consist of arteriovenous malformations and arteriovenous fistulas including SDAVF. Although SDAVF is a rare vascular malformation, it is the most common vascular malformation of the spinal cord and represents approximately 80% of all spinal arteriovenous malformations³⁾. The majority of fistulas affect the thoracolumbar region, and more than 80% of all SDAVFs are located between T6 and L2. Only 2% of patients have SDAVF at the level of the foramen magnum (high cervical lesion). Additionally, low cervical lesions, located below C2 and above T1, are extremely rare⁴⁾. The average annual incidence of SDAVFs is approximately 5-10 cases per one million people. More than 80

Fig. 1. Spinal magnetic resonance imaging findings of spinal dural arteriovenous fistula. (A) T2-weighted image showing extramedullary heterogeneous signal intensity. (B) T2-weighted image 6 months after surgery. It shows no heterogeneous signal intensity.

Fig. 2. A pathologic examination of the arteriovenous malformation. (A) and (B) show a vein with an arterIALIZED, thick collagen coated wall (arrowheads), thrombosis recanalization (arrows), and thrombosis calcification (long arrow). (C) and (D) show an artery with remarkable hyperplasia of smooth muscle, collagen deposition (arrows), reduplication of the internal elastic lamina and dispersal of the elastic membrane (arrowheads). (A: Hematoxylin and eosin stain, $\times 20$. B: Hematoxylin and eosin stain, $\times 20$. C: Elastic stain, $\times 100$. D: Trichrome stain, $\times 100$.)

% of SDAVF patients are men, and two-thirds of patients diagnosed with SDAVF are between 60 and 70 years old⁵. The median age at the time of initial symptoms is 57.6 years⁶. The onset age of our patient was 12-years-old, and the incidence at this age is very low.

SDAVFs are acquired vascular malformations that may be caused by infection, syringomyelia, trauma, and surgery. However, the exact etiology is enigmatic⁵. The pathophysiology of venous hypertension was proposed in 1974 by Aminoff and Logue⁷. For SDAVF, there is typically a feeding radiculo-meningeal artery entering the spinal dura mater close to the spinal nerve root where the arterial blood enters a radicular vein; this radiculo-meningeal artery forms a fistula with the vein. The coronal venous plexus subsequently becomes arterialized. The drainage of arterial blood into the spinal veins through the SDAVF increases spinal venous pressure and decreases the arteriovenous pressure difference. This change decreases the drainage of the normal spinal vein. The obstruction of venous outflow results in chronic venous congestion, venous hypertension, and cord edema. These symptoms lead to chronic medullary ischemia and progressive myelopathy^{4,5}. Hurst et al.⁸ reported that a biopsy from the spinal cord demonstrated the following findings: white matter

with marked hypocellularity, small blood vessels with hyalinized walls, extensive vascular sclerosis, gliosis of the white matter, and focally enlarged degenerating axons. These findings support the pathophysiology of ischemic myelopathy due to increased venous pressure.

The presentation of SDAVF patients is nonspecific. However, SDAVF has a pathognomonic clinical course. The course of SDAVF is typically a slowly progressive myelopathy with symptoms of gait disturbances, sensory loss, bladder incontinence, and erectile dysfunction. According to Jellema et al.⁶, the most common early symptoms of SDAVF were gait disturbances, sensory disturbances and paresthesia. In contrast, the most common symptoms at diagnosis were micturition problems, leg weakness, and sensory disturbances. Our patient's initial symptoms were leg weakness and numbness. A separate case report showed that cauda equina syndrome developed over a relatively short period of 3 months with pure lower motor neuron signs and no upper motor neuron signs⁹. Typically, upper motor neuron signs slowly progress and worsen with SDAVF during a period of 15–23 months^{5,6}.

According to Behrens and Thron¹⁰, the mean time from initial neurological symptom onset to diagnosis of SDAVF was 15.5 months. The diagnosis of SDAVF is difficult because the disease

is rare, and initial symptoms are misleading. The diagnosis can be aided by an MRI scan and catheter angiography, which are required to establish an SDAVF diagnosis. Although myelography was previously used to diagnose SDAVF, MRI scans are now used. MRI findings show swelling of the spinal cord with hypointensities on T1-weighted images and hyperintensities on T2-weighted images. The hypointense “flow void phenomena” at either the ventral or dorsal side of the spinal cord represents the tortuous and enlarged veins¹¹. Specifically, angiography of the spinal cord is the gold standard and enables physicians to identify the exact location of the SDAVF. In our case, an MRI scan was used and led to an early diagnosis and treatment.

The treatment modalities for SDAVF include endovascular embolization and open spinal surgery. During surgery, the intradural shunting vein is disconnected from the perimedullary venous plexus. The disconnection of the arteriovenous shunt eliminates venous congestion and hypertension before irreversible neurological deficit occurs^{4,5}. The results of a meta-analysis performed by Steinmetz et al.¹² suggest that 98% of patients had their fistulas successfully obliterated after the initial treatment with microsurgery. Additionally, 89% of patients showed improvements in neurological symptoms after surgical treatment. Endovascular embolization is reported to have a very low rate of recanalization¹³ and has advantages of being noninvasive. Furthermore, endovascular embolization can lead to immediate angiographic control¹⁴. Our patient underwent a decompression laminectomy without endovascular embolization. The patient's symptoms improved substantially after surgery.

Although no prognostic factors of SDAVF have been reliably established, there are several prognostic factors for functional recovery from SDAVF. These factors include age, duration of symptoms, extent of neurological disability before treatment, and success of fistula closure after the initial treatment. The outcome of SDAVF is generally better in patients presenting with younger age, milder symptoms, or shorter intervals between onset and treatment^{4,5,15}. Some studies indicate that the duration of symptoms before treatment has no direct relation to the post-treatment improvement, but it is certain that patients with more serious symptoms generally have a longer duration of illness¹⁶. The untreated patient runs the risk of an acute deterioration. It is proposed that surgery should not be delayed as each passing month will worsen the eventual outcome⁷.

In this report, we presented the case of a 12-year-old boy with weakness in his upper and lower extremities and alternating hemiplegia. We identified that the patient had a lower cervical SDAVF with an extradural hematoma. This case is clinically important due to the rarity of lower cervical SDAVF in pediatric

patients. There are a limited number of prior studies reporting lower cervical SDAVF in children and adolescents. Several cases of spinal arteriovenous malformation other than SDAVF have been reported^{17,18}. However, to our knowledge, there have been no previous reports of cases involving lower cervical SDAVF with a concurrent extradural hematoma in children and adolescents that were confirmed by histological examination. Physicians may overlook the possibility of SDAVF in children and adolescents because of its nonspecific symptoms and low incidence. In our case, the timely use of an MRI scan facilitated a prompt diagnosis and improved the prognosis by allowing proper immediate treatment. Both pediatricians and pediatric neurologists should be aware of the presentation of SDAVF in pediatric patients to prevent irreversible neurological handicaps associated with misdiagnosis or delay in diagnosis. A delayed diagnosis may lead to worse outcomes and poor treatment responses.

요약

경부 척수 경막 동정맥루(SDAVF)는 진행성 상위 운동 신경원성 증상을 초래하는 드문 혈관 기형으로, 샷길(fistula)은 경추부에서도 발생하지만 대부분은 흉추부와 요추부에 위치한다. SDAVF는 여성에 비해 남성에서의 발생빈도가 4-5배 높고, 중년의 나이에 호발하는 것으로 알려졌으나, 12세 남아에서 초기 증상으로 교대마비를 보이면서 급속히 진행되는 SDAVF 1례를 경험하였기에 이를 보고하고자 한다. 환자의 척수 자기공명영상(MRI)에서 T2 위치에서 신호강도가 감소하였고, C7에서 T2에 걸쳐 척수 배측으로 경막외 종괴 병변이 관찰되고 비균질한 신호강도를 보였다. 증례의 환아는 적절한 시기에 MRI 검사를 시행함으로써 즉각적인 진단을 받을 수 있었고, C7, T1 척추 후궁절개술(laminotomy) 및 T2 감압후궁절제술(decompression laminectomy)을 시행 받은 후 증상이 호전되었다. 조직 검체 병리검사에서 동정맥루에 합당한 소견을 확인하였고, 병리검사 결과를 바탕으로 SDAVF로 최종 진단을 내렸다. 소아에서 SDAVF는 빈도가 매우 낮아 간과될 수 있고, 진단과 치료가 늦어지면 신경학적 결손을 돌이킬 수 없으므로 주의를 요한다.

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