Spinal Dural Arteriovenous Fistula With Lipomyelodysplasia

—Case Report—

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Abstract

A 72-year-old man presented with a very rare case of spinal dural arteriovenous fistula (AVF) with lipomyelodysplasia manifesting as progressive paraparesis and bladder dysfunction. Magnetic resonance imaging revealed a spinal lipoma associated with tethered cord and spinal cord swelling with dilated perimedullary veins. Embolization of the spinal dural AVF was successfully performed, and is an optional treatment for coexisting spinal dural AVF and lipomyelocele in adults.

Key words: spinal dural arteriovenous fistula, lipomyelodysplasia, embolization

Introduction

Lipomyelodysplasia is a common abnormality of the spine. Spinal vascular malformations are rare and can be divided into two categories: congenital lesions, including spinal arteriovenous malformations (AVMs) and cavernomas; and acquired lesions, including dural arteriovenous fistulas (AVFs).1) The coexistence of AVF and lipomyelodysplasia is very rare.1,2,4,7) Here, we report a case of combined treatment of AVF and lipomyelodysplasia by embolization with n-butyl 2-cyanoacrylate (NBCA).

Case Report

A 72-year-old man had been born with a skin-covered soft swelling in the lumbosacral region. Additionally, a dissecting aneurysm of the aorta (DeBakey type IIIb) was diagnosed 4 years before the present admission, and remained stable during the follow-up period. The patient had a 5-month history of progressive fatigue in both legs. No neurological deficit was reported prior to the present admission.

On admission, physical examination identified a brown-red discoloration in the lumbosacral area. Neurological examination demonstrated bladder dysfunction, gait disturbance due to weakness of both lower extremities, and sensory disturbance at levels lower than L3. Lumbar and lumbosacral magnetic resonance (MR) imaging revealed a lipomyelocele containing flow-void structures, indicating AVF, and terminal hydromyelia (Fig. 1A, B). The dilated vessels extended around the spinal cord up to the thoracic level. Spinal cord is tethered by a lipoma with AVF. Diagnostic angiography under general anesthesia was planned to detect the exact location of the fistula. As we had the opportunity, we planned to perform embolization of the fistula after the diagnostic angiography.

Fig. 1 A, B: Preembolization sagittal T1-weighted (A) and T2-weighted (B) magnetic resonance images showing multiple signals around the spinal cord with lipomyelocele, as well as cord tethering and change in cord intensity. C, D: Postembolization sagittal T1-weighted (C) and T2-weighted (D) magnetic resonance images showing that dilated perimedullary vessels had decreased in number in association with improvement in cord intensity.
A microcatheter (Marathon™ Flow Directed Micro Catheter; ev3 Neurovascular, Irvine, California, USA) was inserted into the second right lumbar artery via the 4 Fr. Cobra catheter. NBCA (17%, 0.5 ml) was slowly injected. After removing the microcatheter, contrast medium was injected from the second right and third right lumbar arteries, but the AVF was not observed (Fig. 2D, E). All procedures were completed without complications. Three months after the embolization, MR imaging demonstrated that the dilated perimedullary vessels had decreased in number and the cord intensity had improved (Fig. 1C, D). Thereafter, the progressive fatigue in both legs disappeared gradually.

Discussion

Spinal lipomas are thought to consist of three subtypes: intramedullary lipomas, lipomyelomeningoceles affecting the spinal cord, and lipomas of the filum terminale with limited involvement of the lowest level of the conus medullaris.1,8 This mesenchyme could prevent closure of the neural tube focally. Spinal AVMs are of congenital origin, whereas dural AVFs are considered to be acquired lesions, occurring in patients over 40 years of age.31 Only four cases of combined spinal lipoma and spinal dural AVFs have been reported.1,2,4,7 One of the AVFs was combined with sacral filum terminale lipoma, whereas the other three were located within a lumbar lipomyelomeningocele located intraspinally or extraspinally. The main features of spinal dural AVFs are late and gradual presentation of symptoms, and lack of association with other vascular or non-vascular malformations, which suggest an acquired process. The coexistence of these two lesions could be due to abnormal angiogenesis in the lipomatous tissue proximal to a dural defect, a site of potential arteriovenous communication.

The majority of patients with lipoma are neurologically intact at birth, but progressive neurological deterioration develops at an early age.31 Late presentation, as in our case, is very rare. However, two previous patients with combined spinal lipomyelomeningocele and spinal vascular malformation also presented late in life.4,6 Gradually, progressing motor weakness and bladder dysfunction were the main neurological problems observed in our patient. These symptoms could develop due to three possible mechanisms: the mass effect of the tumor; tethering of the cord because of the lipoma; and chronic venous hypertension from the AVF.31 Angiography of ischemic spinal cord due to tethered cord or venous hypertension can cause abrupt exacerbation of the symptoms. Venous hypertension is the most likely pathogenic mechanism. Lipoma attached to the “cul de sac” of the dura mater (a site of potential arteriovenous communication) may have produced local hypervascularization with subsequently acquired AVF.30 Another possibility was that both the potential arteriovenous shunt and mechanical stimulation by the lipoma or spinal cord tethered to the dura mater led to the venous hypertension. According to the condition of our patient, extended surgery had higher risks than embolization followed by diagnostic angiography. Therefore, we performed only embolization to improve the perfusion of the conus by reducing the venous congestion secondary to the AVF.

In conclusion, although very rare, lipomyelomeningocele can be associated with spinal dural AVF. Embolization of the spinal dural AVF can be an optional treatment for coexisting lipomyelomeningocele and spinal dural AVF in adults.
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Conflicts of Interest Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices in the article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

References


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