

Dural Arteriovenous Fistula at the Foramen Magnum with Holocord Myelopathy: Case Report

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We report a case of dural arteriovenous fistula at the foramen magnum with a diagnostic delay of 10 months from initial myelopathic symptoms. A 59-year-old male with urinary incontinence and progressive ascending weakness of the extremities was transferred through two other hospitals to our institution with no tentative or established diagnosis. A correct diagnosis was not made until 10 months after initial symptoms: follow-up MR images revealed diffuse edema involving the holocord and lower brain stem, and engorged veins around the spinal cord. Cerebral angiography confirmed a dural arteriovenous fistula drained by perimedullary venous plexus. That fistula was occluded by transarterial embolization using NBCA (n-butyl cyanoacrylate). However, it seemed hard to expect any improvement in the clinical condition of the patient due to considerable diagnostic delay. To avoid missing a chance of treatment for this potentially reversible disease, early diagnosis is mandatory and it requires a high-level suspicion.

Key Words : Dural arteriovenous fistula; Foramen magnum; Perimedullary venous drainage; Myelopathy

Dural AVF at the foramen magnum with perimedullary venous drainage is known to cause slowly progressive ascending myelopathy involving first the lower limbs and then the upper limbs. Often sphincter disturbances are present (1, 2). Even though it cannot provide precise location of fistula, MRI appearance is virtually pathognomonic. A swollen cord with central myelopathy and engorged perimedullary veins are indicative of the diagnosis of a fistula, either cervical spinal dural AVF or an intracranial dural AVF with perimedullary venous drainage (3). The case of this study had typical but rare symptoms and imaging findings, so that a correct diagnosis was delayed to

irreversible cord injury. Little improvement was expected despite curative embolization of the fistula in this patient.

CASE REPORT

A 59-year-old male presented with paraparesis 10 months ago. When he visited one of the hospitals in the local area of his hometown, spine MR images were taken and demonstrated multisegmental edema in the cervical cord and conus medullaris region. With these findings, transverse myelitis was diagnosed and the patient was transferred to another hospital to treat transverse myelitis. Based on our medical record, other clinical conditions could not be achieved in detail. However, other findings toward a correct diagnosis were missed in those hospitals: abnormal enhanced vascular structures around the spinal cord on contrast-enhanced T1-weighted images and enlarged and tortuous perimedullary vessels in the cervical region on contrast-enhanced MR angiography (Fig. 1). Six months after the first diagnosis of transverse myelitis,

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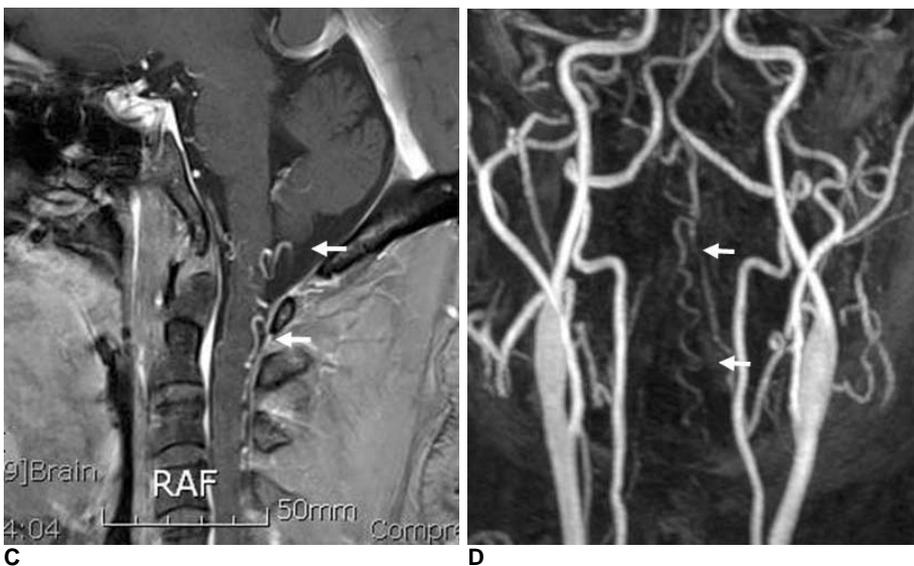
he deteriorated to quadriplegia with some fluctuation and eventually to quadriplegia at 10 months. With 10-month-delay of diagnosis, he was transferred to our

institution.

Neurologic examination on admission revealed complete quadriplegia, hypesthesia under the T7 level,



Fig. 1. MR images taken just after initial symptoms.
A. T2-weighted image shows multisegmental intramedullary hyperintensities in the cervical cord and around the conus medullaris with suspicious multiple flow voids (arrows).
B. Abnormal enhanced vascular structures were demonstrated on contrast-enhanced MR image around the spinal cord (arrows).
C. Craniocervical contrast-enhanced T1-weighted image shows abnormal enhanced vascular structures at the level of the foramen magnum (arrows).
D. Contrast-enhanced MR angiogram reveals an enlarged tortuous perimedullary vessel in the midline cervical region (arrows) suggesting the presence of a vascular lesion.



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Fig. 2. 10-month follow-up MR images.

A, B. T2-weighted spine MR images show swelling and hyperintensity of the holocord and lower brain stem.
C. Patchy and diffuse enhancement of the spinal cord is shown on contrast-enhanced T1-weighted MR image.

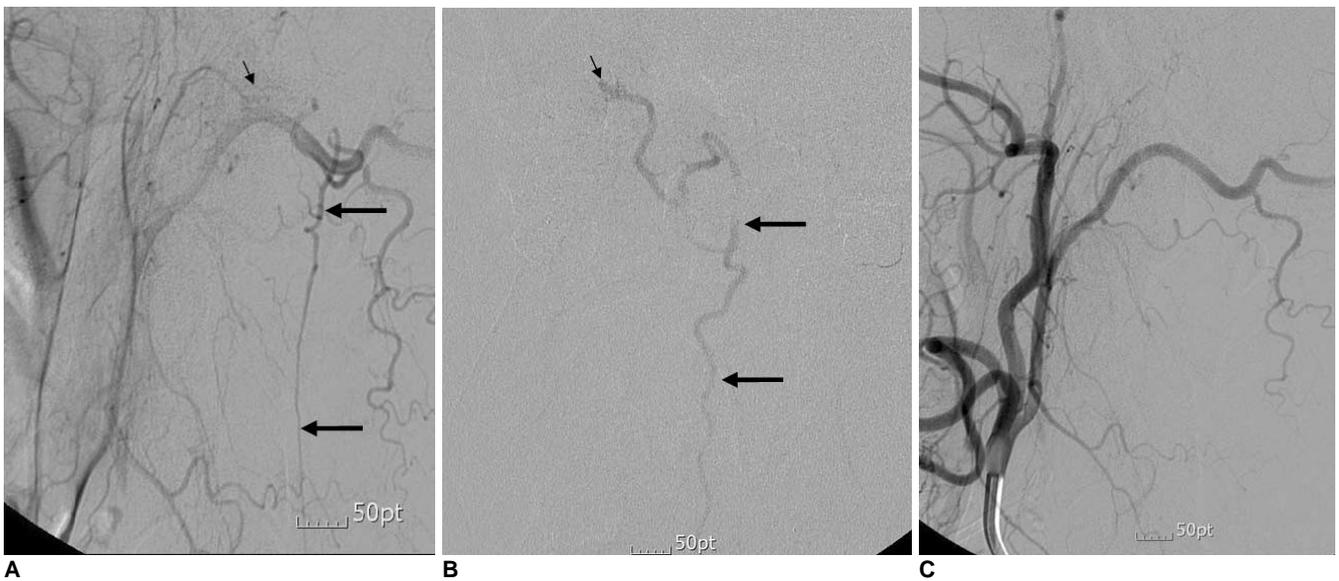


Fig. 3. Digital subtraction angiography of the right external carotid artery.

A. Lateral projection shows dural arteriovenous fistula at the foramen magnum (short arrow) and draining perimedullary vein (long arrows).
B. Frontal projection of superselective angiography of the hypoglossal branch of the right ascending pharyngeal artery shows a short fistulous segment (short arrow) and draining midline perimedullary vein (long arrows). This perimedullary vein is the same in shape and location as the one on contrast-enhanced MR angiogram (1D).
C. Lateral projection of completion angiography postembolization shows no residual fistula and draining perimedullary vein.

and urinary incontinence. He was in status of colostomy due to fecal incontinence in another hospital. Follow-up spine MR images demonstrated diffuse edema and patch enhancement of the holocord and lower brain stem (Fig. 2). Cerebral angiography was performed and revealed a dural AVF at the foramen magnum supplied by only hypoglossal branch of ascending pharyngeal artery and drained into the anterior and posterior perimedullary veins (Fig. 3A and 3B). To obliterate the fistula, superselective embolization of the feeding artery was performed using 25% N-butyl cyanoacrylate (NBCA). The fistula and venous drainage into the perimedullary vein disappeared after embolization (Fig. 3C). Two days after embolization, the patient was transferred to another rehabilitation center with no change of neurologic deficits.

DISCUSSION

Dural arteriovenous fistula (AVF) at the foramen magnum with perimedullary venous drainage is not a common disease entity (4). The fistula is supplied by meningeal branches of the vertebral artery and/or external carotid artery, and the venous drainage comes out intradurally into the perimedullary venous plexus, resulting in venous hypertension that is responsible for the myelopathic symptoms (5). That has been classified as type V dural AVF in the classification scheme proposed by Cognard et al. (6), wherein progressive myelopathy developed in 50% of cases.

Typical MR imaging findings are known as spinal cord edema and engorged perimedullary vessels. Symptomwise, progressive ascending motor and sensory symptoms also suggest dural AVF at the foramen magnum or intracranial dural AVF with perimedullary venous drainage. However, scarcity of these lesions and the discrepancy between neurological or topographical findings and location of the AVF are likely to delay a correct diagnosis of this disease in daily practice.

Our patient referred to our institution with a 10-month delay after the initial onset of his symptoms. Although the spine MRI and contrast-enhanced MRA

taken in the first hospital demonstrated the hallmarks of a spinal vascular lesion, the patient was diagnosed as having transverse myelitis and treated with conservative management despite progressive clinical deterioration for 10 months. The diagnostic and subsequent therapeutic delay was responsible for the permanent neurologic deficit in the patient. Even paraplegia might be reversible if the fistula is diagnosed and treated early enough before ischemic and gliotic changes have been irreversible (7).

In conclusion, dural AVF should be considered first in a patient with myelopathic symptoms and MR imaging findings typical of a spinal vascular lesion regardless of lesion location. Earlier diagnosis could be made with a high index of suspicion as well as thorough understanding of clinical and imaging findings.

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전척수 척수병증을 동반한 대후두공 경막동정맥루

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경막동정맥루는 모호한 증상 및 영상 소견으로 인하여 진단이 어려운 경우가 있다. 첫 증상 발생 후 10개월만에 진단된 후대뇌공의 경막동정맥루의 증례를 보고한다. 59세 남자환자가 요실금과 진행되는 상행성 사지 근력약화로 두 곳의 타기관에서 적절한 진단 없이 본원으로 전원되었다. 증상이 발생한 이후 10개월 동안 잘못된 진단 하에 치료를 받았으며, 10개월 추적 자기공명영상에서 전척수 및 하부 뇌간에 걸쳐 미만성 척수 부종과 척수 주변에 충혈된 정맥들이 관찰되었다. 뇌혈관 조영술을 시행하여 척수주위 정맥총으로 유출되는 대후두공 경막동정맥루를 확진하였다. 이 동정맥루는 경동맥접근법을 통해 NBCA를 이용해 색전술을 시행하였다. 그러나 상당히 오랜 기간동안 진단이 지연된 것으로 인해 환자의 임상적 호전은 기대하기 어려웠다. 이러한 경막동정맥루와 같이 치료에 의해 가역적인 질환은 영구적 손상이 오기 전 치료시기를 놓치지 않아야 하며, 이를 위해 조기 진단이 필수적이다.

Key Words : Dural arteriovenous fistula; Foramen magnum; Perimedullary venous drainage; Myelopathy