A Case of Dural Arteriovenous Fistulas at the Craniocervical Junction Presenting with Occipital/Neck Pain Associated with Sleep

Satoshi Suda, Ken-ichiro Katsura, Seiji Okubo, Arata Abe, Takuya Kanamaru, Masayuki Ueda, Masahiro Mishina, Mineo Yamazaki and Yasuo Katayama

Abstract

Venous congestive myelopathy associated with spinal dural arteriovenous fistulas (DAVFs) is a treatable disorder that can be controlled without sequelae if it is diagnosed at an early stage. It is important to consider spinal DAVFs in the differential diagnosis based on clinical history and neurologic examination. We report the unique case of a patient with DAVFs at the craniocervical junction presenting with occipital/neck pain associated with sleep.

Key words: secondary headache disorders, occipital/neck pain, spinal dural arteriovenous fistulas, sleep

(Intern Med 51: 925-928, 2012)
(DOI: 10.2169/internalmedicine.51.6607)

Introduction

Spinal dural arteriovenous fistulas (DAVFs) are acquired arteriovenous shunts located within the spinal dura; they commonly occur at the thoracolumbar region in middle-aged men. Presenting symptoms and signs include slowly progressive venous congestive myelopathy with paraparesis, sensory impairment of the lower extremities, and bowel and bladder dysfunction (1, 2). Only 2-5% of patients with spinal DAVFs show craniocervical junction involvement (3, 4). The rare and nonspecific clinical presentation of such DAVFs may lead to delayed diagnosis and treatment, resulting in irreversible neurologic damage. We report the case of a patient with DAVFs at the craniocervical junction presenting with occipital/neck pain associated with sleep.

Case Report

A 76-year-old man with obstructive sleep apnea syndrome (OSAS) and congestive heart failure (CHF) was admitted to our hospital with sudden-onset paraplegia. He reported awakening at 2-3 AM (after sleeping for approximately 4 hours) with occipital/neck pain every night for 10 days before admission. The pain was sometimes described as “throbbing” and accompanied by nausea, but he experienced no visual or autonomic symptoms. His pain was moderately resolved after sitting for a few hours, and he experienced only mild pain later in the day. He was diagnosed with degenerative cervical disk disease by his local doctor and treated with analgesics, but the pain did not decrease. On admission, neurologic examination revealed mild dysarthria, complete flaccid paraplegia, impaired sensation, and vesicorectal dysfunction. T2-weighted magnetic resonance imaging of the brain and cervical spinal cord showed an abnormal hyperintense area extending from the left side of the medulla to C7, which was associated with cord swelling and perimedullary flow voids (Fig. 1, 2). His symptoms progressed rapidly thereafter: he experienced severe muscle weakness in the upper extremities, and respiratory function became impaired 2 days after admission. Left vertebral angiography demonstrated DAVFs fed by the meningeal branch of the vertebral artery (Fig. 3A-D). Surgical treatment was impossible because of his rapidly worsening systemic conditions associated with CHF and aspiration pneumonia. Guglielmi detachable coil embolization of the vertebral artery was performed, because it was difficult and dangerous to insert a microcatheter into the feeding artery. No abnormal
Figuress 1. Axial diffusion-weighted (A) and T2-weighted (B) brain magnetic resonance images demonstrating mild abnormal hyperintensity in the left medulla. The fluid-attenuated inversion recovery sequence more clearly shows the hyperintensity, which is compatible with vasogenic edema (C).

Figure 2. Sagittal (left) and axial (right) T2-weighted magnetic resonance images of the cervical spine, showing mild cervical disk degeneration and an abnormal hyperintense area extending from C1 to C7 associated with cord swelling and abnormal flow voids along the surface of the spinal cord.

Discussion

Spinal DAVFs are difficult to diagnose, and treatment is often delayed after the first symptoms appear. Since outcomes are worse with more severe neurologic deficits, it is important to consider spinal DAVFs in the differential diagnosis from the beginning, and appropriate radiologic examinations should be conducted promptly. The first symptom in the present patient was occipital/neck pain during sleep, with no obvious signs of myelopathy. The initial symptoms of spinal DAVFs typically include gait difficulties and sensory symptoms such as paresthesia and sensory loss, but also back pain possibly resulting from compression of the adjacent nerve root or traction of the dura (1, 2). These findings and the present patient’s dramatic improvement after coil embolization likely indicate that the DAVFs at the craniocervical junction in our patient were related to the new-onset refractory occipital/neck pain experienced during sleep.

Venous congestion with intramedullary edema is considered to be a causative mechanism of neurologic deterioration in spinal DAVFs (5, 6). Some reports have described patients with thoracolumbar spinal DAVFs who typically show slowly progressing paraparesis that may be acutely or subacutely exacerbated after physical activity, rapid infusion of methylprednisolone diluted in saline solution, or lumbar puncture, and may be secondarily caused by a further increase in venous hypertension (7-9). While sleeping in the supine position, the present patient developed severe pain, which was alleviated after sitting for a few hours, and he only had mild pain during the day.

A possible explanation for the presentation of the present patient is that the increased venous pressure caused by CHF and OSAS while sleeping in the supine position may have resulted in occipital/neck pain (10, 11). Moreover, venous pressure in the neck, where the DAVFs were located in the
Figure 3. Lateral views (A, early phase; B, late phase) and anteroposterior views (C, early phase; D, late phase) of left VA angiograms revealing spinal dural arteriovenous fistulas supplied by a radicular artery from C1 and draining via a spinal vein to the lower cervical level. Post-embolization angiograms (E, right VA angiogram; F, left VA angiogram) demonstrate complete disappearance of the abnormal vessels.

The present patient, is generally approximately 20-40 mm Hg lower than the aortic pressure in the upright position, which is caused by gravity; in contrast, venous pressure in the thoracolumbar region, where spinal DAVFs are usually located, is generally approximately 20-40 mm Hg higher than the aortic pressure in the upright position. In the supine position, however, venous pressure in the neck can increase to the same level as that in the heart (12). Although this report involves a single case description and the precise mechanism underlying this patient’s clinical presentation remains unclear. We speculate that the occipital/neck pain that developed during sleep in our patient may have been caused by the increase in venous pressure related to his coexisting diseases and the position of the spinal DAVFs.

Occipital/neck pain is nonspecific and constitutes a common problem, especially in the elderly. This case illustrates that DAVFs at the cranio cervical junction can be considered as one of the differential diagnosis of occipital/neck pain. Our observations, combined with previous reports, may suggest that changes in symptoms based on hemodynamic alterations may lead to an earlier diagnosis of spinal DAVFs.

The authors state that they have no Conflict of Interest (COI).

References

8. Khurana VG, Perez-Terzic CM, Petersen RC, Krauss WE. Singing paraplegia: a distinctive manifestation of a spinal dural arte-

