MR Findings of Subacute Necrotizing Myelopathy: Case Report

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Purpose: Subacute necrotizing myelopathy (SNM) is a rare non-tumorous disease of spinal cord characterized by subacute clinical course of progressive neurological deterioration. We report MR findings of a patient with pathologically proved SNM.

Materials and Methods: 1 case of pathologically proved subacute necrotizing myelopathy

Results: The patient was a 56-year-old man with progressive motor weakness and sensory loss of the lower extremities, and urinary and fecal incontinence for 11 months. Spine MRI revealed diffuse enlargement of the thoracic spinal cord from T2 to T7 level. Signal intensity of the expanded spinal cord was isointense relative to normal cord on T1-weighted image and hyperintense on proton-density and T2-weighted images. On contrast enhanced T1-weighted image, there was diffuse homogeneous enhancement in the expanded cord lesion.

Conclusion: MR demonstration of stable persistence of spinal cord lesion or atrophy over months or years with clinical findings of gradual progressive neurologic deterioration may be helpful in the diagnosis of SNM(1).

Index Words: Thoracic spinal cord, MR Thoracic spinal cord, myelitis

Subacute necrotizing myelopathy (SNM) or Foix-Alajouanine disease is an uncommon disease that is clinically characterized by subacute progressive neurological deterioration and pathologically by spinal cord necrosis. The underlying pathophysiology of SNM has been believed to be venous hypertension caused by spinal dural arteriovenous fistula (AVF). However, the etiology has been the subject of controversy(1, 2). Preoperative diagnosis of SNM is difficult unless a spinal dural AVF is demonstrated by angiography, and confirmative diagnosis is usually made by biopsy. MRI findings of SNM have been documented in a few reports(1, 2). We report MRI findings of a biopsy-proved case.

CASE REPORT

A 56-year-old man has suffered from progressive weakness and sensory loss of the lower extremities and urinary and bowel dysfunction for 11 months. He was healthy until May 1992, when weakness in his right leg developed. The symptom has progressed slowly. In January 1993, weakness in his left leg occurred, and the patient could no longer rise or walk. Two months later, he developed sensory loss in both lower extremities and fecal incontinence and urinary dysfunction. At the time of admission in April 1993, there was a flaccid paraplegia. On neurological examination, deep tendon reflexes were absent in both legs, while preserved in the arms. The sensation of pain, temperature, vibration, position was impaired below T4 level and there was complete loss of sensation below T6 level. Cerebrospinal fluid (CSF) analysis showed that protein levels were elevated up to 3620 ng/dl. Electromyography (EMG) and nerve conduction velocity study suggested diffuse myelopathy involving anterior horn cells. Tumor marker and basal hormone studies were normal. Initial MR imaging of cervico-
thoracic spine obtained on April 15, 1993, revealed diffuse enlargement of the thoracic spinal cord from T2 through T7. The expanded cord lesion showed isointense signal on T1-weighted image, and hyperintense signal on proton-density and T2-weighted images. On contrast enhanced T1-weighted image, there was diffuse homogeneous enhancement of the expanded cord lesion at T3–T6 levels (Fig. 1). There was no evidence of abnormal vascular structures suggesting spinal dural AVF. The patient underwent surgery with preoperative diagnosis of intramedullary tumor. At surgery, there was diffuse cord swelling and engorged veins on the dorsal surface of the cord, but no dural AVF was found. On the microscopic examination of pathologic specimen, there were abnormal vascular channels in the background of necrotic tissue and gliosis, highly suggesting SNM. After operation, steroid therapy was done but the paraplegia persisted without improvement. Follow-up MRI obtained 2 months after operation showed persistent diffuse swelling of thoracic spinal cord with contrast enhancement at T8–T11 levels.

DISCUSSION

In 1926, Foix and Alajouanine described two patients with subacute myelopathy, in whom abnormal dilated tortuous vessels on spinal cord surface were revealed by postmortem examination. Later on, the association of this disease with arteriovenous malformation (AVM) of spinal cord has been reported, and Foix-Alajouanne disease was suggested to be a complication of spinal AVM that was caused by thrombosis within the abnormal vessels of the spinal cord (3, 4).

However, the underlying pathophysiology of Foix-Alajouanne disease is now believed to be spinal venous hypertension secondary to spinal dural AVF. It is widely thought that spinal dural AVF draining into the spinal medullary vein results in venous hypertension and leads to progressive intramedullary congestion and cord ischemia (5, 6, 7). The nidus of dural AVF is embedded in dura covering the proximal nerve root.

Fig. 1. T1-weighted image(a) shows diffuse enlargement of the thoracic spinal cord from the level of T2 through T7 and isointense signal of the lesion. T2-weighted image(b) shows hyperintense signal. On contrast enhanced T1-weighted image(c), there is diffuse homogeneous enhancement of the expanded cord lesion. At surgical biopsy, it was proved to be subacute necrotizing myelopathy.

Fig. 2. Characteristic endothelial proliferation forming multiple vascular lumina in the background of ischemic necrosis and associated gliosis.
and in the adjacent spinal dura. Most of the spinal dural AVFs are found at or below the midthoracic level. This disease is more common in aged men. There is initial spastic paraparesis followed by flaccid paraplegia. Sensory deficit or paresthesia is almost always found, and bowel or bladder dysfunction may ensue. The prognosis has not been well documented, but the reported prognosis was dismal(4, 7). Early obliteration of the dural AVF may eliminate the cause of venous congestion and stasis, prior to irreversible cord damage(4). After irreversible cord damage, however, significant improvement seems to be impossible. This disease usually does not respond to steroid therapy(1). The present case showed the clinical course and the pathologic findings similar to Foix-Alajouanine disease, which is pathologically characterized by abnormal intramedullary vascular channels in the background of gliosis and necrosis. Because the myelography and spinal angiography were not performed in the present case, the presence of spinal dural AVF was not completely excluded. Thus, in strict sense, our case does not meet the classic Foix-Alajouanine disease. However, no demonstration of abnormal vascular structures suggesting dural AVF on both MRI and operative findings strongly suggests the absence of spinal dural AVF. It remains controversial whether the SNM which is a pathologic finding of Foix-Alajouanine disease is always associated with spinal dural AVF or not (1, 2).

Mirich et al(1) reported MR findings of pathologically proved 4 cases of SNM which had no identified underlying spinal dural AVF. The initial MR findings showed focal cord enlargement in all 4 cases. Follow-up MRI showed cord atrophy in one case, further slight enlargement of cord in two cases, and no change in remaining one case. The lesions showed nonspecific T1 and T2 lengthening of the signal intensity on MR images in all cases and peripheral rim enhancement on contrast enhanced T1 weighted image in only one case. Meanwhile, Larsson et al(8) described MR findings of venous infarct of spinal cord in 3 cases with spinal dural AVF presented with acute or progressive myelopathy. The MR images showed focal enlargement of spinal cord in two cases and focal atrophy in one case at thoracic level. There were T1 and T2 lengthening of the signal intensity and diffuse enhancement of the cord lesion in all cases. In one case, MR image showed intrathecal serpentine vascular abnormalities. In another case, persistent cord enhancement was seen on 1 year follow-up study. Even though these 3 cases were not pathologically proved to be SNM, the cord lesions of these cases are thought to have the same pathology considering the documented pathophysiology of SNM. The initial MR imaging of the present case showed MR findings similar to those of previous cases described by Mirich et al(1). Contrast enhanced T1-weighted image showed nonspecific diffuse enhancement in the expanded cord, unlike rim enhancement described by Mirich et al(1). This suggests that the enhancement pattern of SNM is variable. As seen in the present case and previous cases reported by Mirich et al(1), SNM may not be associated with spinal dural AVF.

Clinically and radiologically, it seems very difficult to differentiate SNM from primary intramedullary cord tumor or transverse myelitis(1, 2). And preoperative diagnosis of SNM is difficult to determine unless a spinal dural AVF is demonstrated by radiologic study. Only MR imaging findings of SNM including signal intensity and enhancement pattern have not so much value for the differential diagnosis unless a spinal dural AVF is demonstrated. MR demonstration of stable persistence of spinal cord lesion or atrophy over months or years with clinical findings of gradual progressive neurologic deterioration may be helpful in the diagnosis of SNM(1).

REFERENCES

아급성 괴사성 척수증(Subacute necrotizing myelopathy)의 자기공명영상(MRI) 소견: 증례 보고

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목 적: 아급성 괴사성 척수증 혹은 Foix-Alajouanine 병은 임상적으로는 아급성 신경 장애를, 병리학적으로는 척수의 괴사 손상이 특징적으로 나타내는 드문 질환으로서, 척수의 경막 동정맥루에 의한 이차적으로 생긴 척수정맥 고혈압이 원인일 것으로 알려져 있으나 아직 확실히 밝혀진 바 없다. 저자들은 수술 후 생검을 통해 진단된 아급성 괴사성 척수증 1예의 MRI 소견을 보고하는 바이다.

대상 및 방법: 수술 및 병리조직학적으로 진단된 아급성 괴사성 척수증 1예

결 과: 환자는 56세 남자로서 점진적인 하지 마비와 감각 소실 그리고 뇌실금과 뇌실금의 증상이 있었고 수술적 생검 결과 병리학적 소견으로는 아급성 괴사성 척수증을 시사하는 척수내 비정상 혈관과 척수의 괴사 및 신경교증의 소견이 있었다. 그러나 MR 소견 및 수술 소견의 경막 동정맥루는 발견되지 않았다. MR 소견은 흉수 부위의 미만성 팽창이 T2-T7에 걸쳐 있었고 병변 부위는 T1 강조영상에서 정상 흉수와 비슷한 신호강도를 보였고 T2 강조영상 및 양성자 강조영상에서는 고신호 강도를 나타냈다. 또한 미만성 팽창을 보이는 병변 부위는 조영증강 후 미만성으로 균일하게 증가된 신호강도를 나타냈다.

결 론: 아급성 괴사성 척수증의 자기공명영상 소견은 비특이적이며 임상적으로 아급성 신경 장애를 보이는 환자에서 자기공명영상에서 수개월 혹은 수년에 걸친 지속적인 척수의 병변이나 척수의 위축이 관찰될 때 아급성 괴사성 척수증의 진단에 도움이 되리라 생각된다.