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J Korean Neurosurg Soc 42 : 64-66, 2007

Case Report

Jae-Ik Cho, M.D.
Young-Dae Cho, M.D.
Young-Don Kim, M.D.

Multiple Spinal Intramedullary Cavernous Malformation with Multiple Intracranial Involvement

Intraspinal cavernous malformation (CM) accounts for 5% to 16% of all spinal vascular abnormalities. Multiple spinal cord CMs are very rare and only a few cases have been described. We report a patient presented with right chest paresthesia and seizure, and diagnosed as multiple spinal intramedullary CM and intracranial involvement.

KEY WORDS : Cavernous malformation · Intramedullary · Multiple · Spine.

Department of Neurosurgery
Daegu Catholic University
School of Medicine
Daegu, Korea

INTRODUCTION

Cavernous malformation (CM) is uncommon central nervous system (CNS) vascular disorder[18], and constitutes 9% of CNS vascular malformations[2]. These angiographically occult lesions are well-circumscribed entities, consisting of thin-walled, lobulated vascular channels, without intervening neural tissue[5]. The spinal cord is an uncommon site for CMs, and when they do occur, are frequently in the epidural space; intramedullary ones are rare[1,11]. Multiple spinal intramedullary CMs are very rare, and only a few cases have been described. We report a case of multiple spinal intramedullary CM with multiple intracranial involvement.

CASE REPORT

A 33-year-old male with right-sided chest tightness and tingling sensation was referred to our hospital for seizure. This was the first occurrence of a generalized tonic-clonic seizure. He had no prior history of seizure, trauma, or febrile convulsion. Band-like right chest sensory changes had begun 2 months before. Physical and neurological examinations were normal, except for hypesthesia at the right T6,7 dermatome level. There were no abnormal laboratory findings and interictal electroencephalogram was normal.

Brain magnetic resonance imaging (MRI) revealed multiple CMs on the cerebral hemisphere, cerebellum, and brain stem (Fig. 1). The right temporal lobe CM was the largest and was considered to be the seizure focus.

Spine MRI showed two hyperintense lesions with dark signal rim on T2 weighted images and iso-to-hyperintense on T1 weighted images at C2-3 and T5 levels. They were not contrast-enhancing (Fig. 2, 3).

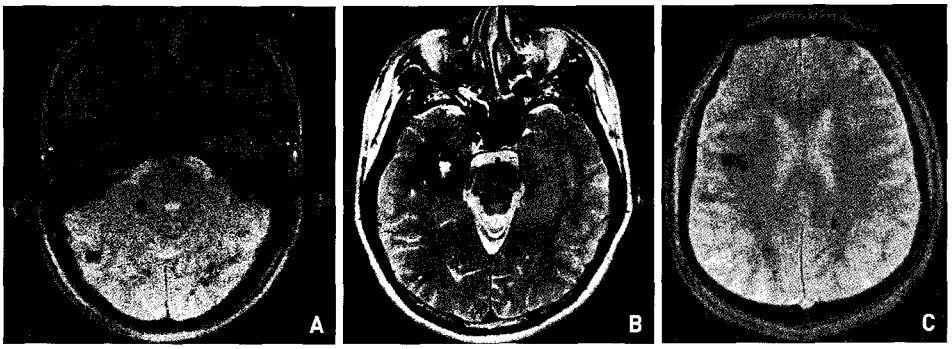

Fig. 1. Axial magnetic resonance images revealing multiple intracranial cavernous malformation in the brain stem, cerebellum (A), right temporal lobe (B), and cerebral hemisphere (C).

• Received : May 2, 2007
• Accepted : June 25, 2007
• Address for reprints :
Young-Dae Cho, M.D.
Department of Neurosurgery
Daegu Catholic University
School of Medicine
3056-6 Daemyeong 4-dong
Nam-gu, Daegu 705-718, Korea
Tel : +82-53-650-4253
Fax : +82-53-650-4932
E-mail : aronnn@empal.com

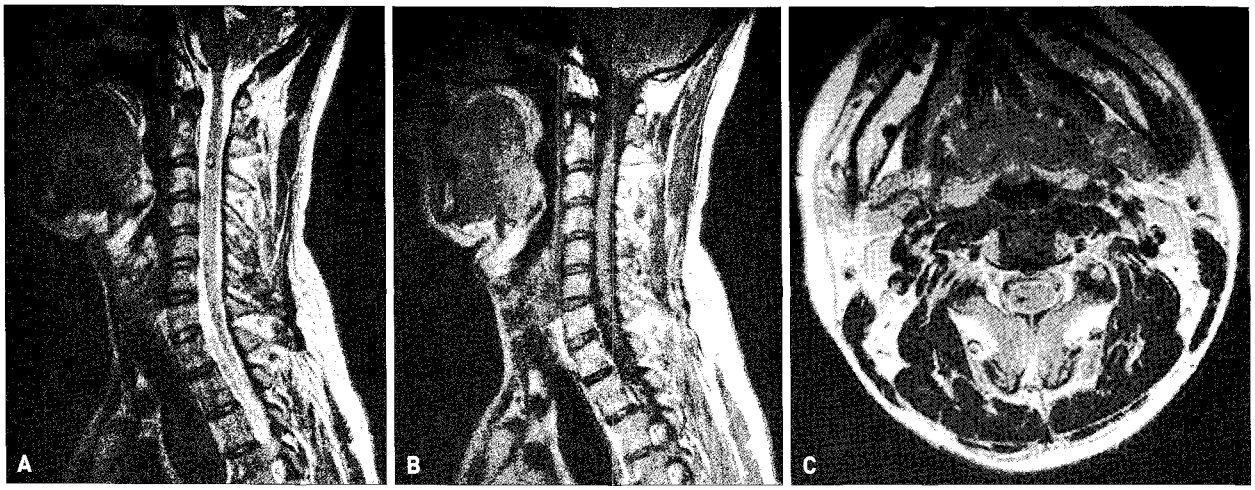

Fig. 2. Cervical magnetic resonance imaging T2-(A), T1-(B) weighted sagittal and T2 weighted axial (C) sections showing a intramedullary lesion at the level of C2-3.

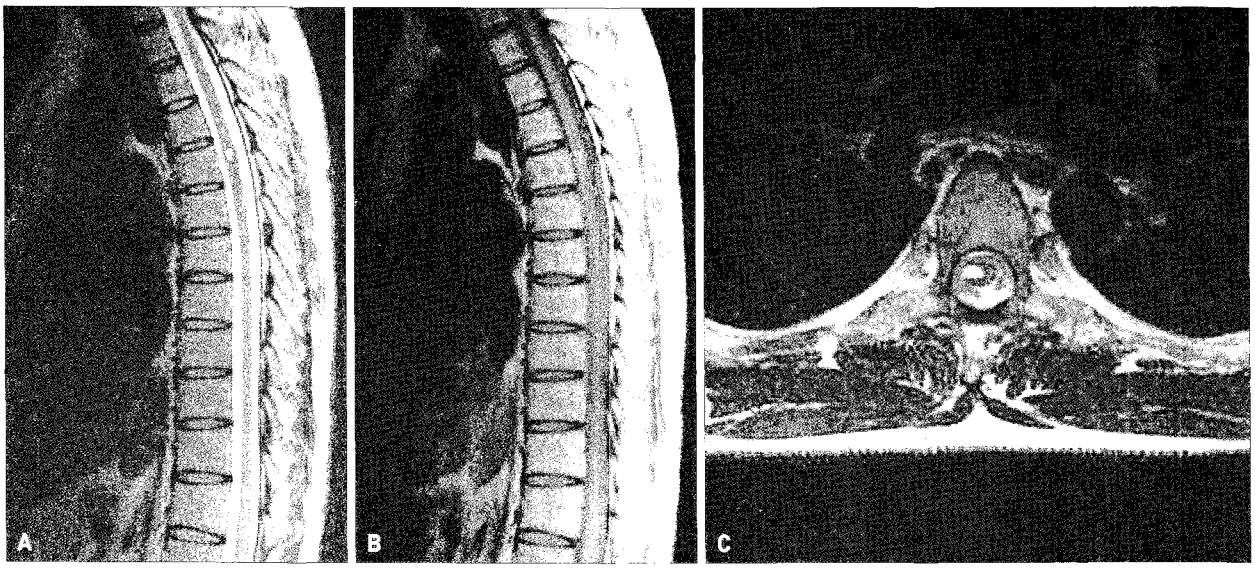

Fig. 3. Thoracic magnetic resonance imaging T2-(A), T1-(B) weighted sagittal and T2 weighted axial (C) sections showing a intramedullary lesion at the level of T5.

Previous patient family history did not have any evidence of neurological disease. But, his daughter's screening MRI in the neuraxis showed multiple intracranial cavernous malformation without spinal involvement.

Patient refused operation of intracranial and thoracic CM, and was discharged with symptomatic improvement after medical treatment.

DISCUSSION

Spinal cord CMs can be extradural, intradural extramedullary, or intramedullary[9,10,19]. Lesions are most frequently localized at the cervical and thoracic spinal cord, but may be seen at any level from the upper cervical cord to cauda equina[1,22]. They may be asymptomatic, or they may cause pain, myelopathy, or sensorimotor deficit due to hemorrhage and mass effect[15,20]. Widespread use of MRI has resulted in enhanced sensitivity and specificity for CM diagnosis[6,11,14,16].

Intraspinal CMs account for 5% to 16% of all spinal vascular abnormalities[4,7,16,19,21]. Multiple spinal cord CMs are very rare, and only a few cases have been described. Zevgaridis et al.[25] reviewed 116 patients with intramedullary spinal CM which had been published between 1903 and 1996. They found only one patient who had two spinal intramedullary CMs. Vishteh et al.[22] reported 17 patient with intramedullary spinal cord CM. Only one patient had a multiple CM in the spinal cord.

Intraspinal CMs commonly accompany intracranial CMs[22,23]. Coexistence of CMs in the brain and spinal cord typically occurs in patients with the familial form of CM[3,17,24]. It has an autosomal dominant pattern of inheritance with incomplete penetrance[12]. Genetic analysis has identified

foci on chromosomes 7q11-21, 7p13-15, and 3q25, 2-27[8,13]. Cohen-Gadol et al.[5] reported that as many as 40% of patients with a spinal CM may harbor a similar intracranial lesion, and approximately 40% with coexisting spinal and intracranial CMs may have the nonfamilial (sporadic) form.

Our patient has the familial form of multiple spinal intramedullary CMs, located in the cervical and thoracic spinal cord, with multiple intracranial involvement. We consider that it is necessary to study genetical analysis of him and his family.

CONCLUSION

A case of multiple spinal intramedullary CM with multiple intracranial involvement is presented. It deserves an attention due to its rarity and is an important reminder to search the neuraxis (brain and whole spinal canal) in patients with spinal intramedullary CM, even if asymptomatic.

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