CASE REPORT

J Korean Neurosurg Soc 40 : 273-276, 2006

Spinal Cavernous Hemangioma Causing Sudden Paraplegia in a 23-Month-Old Kid

Jae Hoon Cho, M.D., You-Nam Chung, M.D., Kyu-Chang Wang, M.D., Byung-Kyu Cho, M.D.

Department of Neurosurgery, Seoul National University College of Medicine, Seoul National University Hospital, Seoul, Korea

Although cavernous angioma itself is not rare, the epidural spinal localization is uncommon and makes preoperative differential diagnosis difficult. An extraordinary case of a thoracic epidural cavernous angioma in very young age, causing sudden paraplegia is presented. Only 79 cases have been reported in the literatures and among them, this kid was the youngest. A 23-month-old boy was referred to us with a 2-day history of sudden both lower limb weakness. Two days before admission, he got up at morning and was unable to stand and even to move the legs. MRI revealed an epidural mass surrounding spinal cord associated with cord compression at the level of the C5 through T3. Through posterior approach with exposure of C6 to T3 level, the hematomatous mass was removed subtotally due to intraoperative bleeding and its ventral location. After the first operation, the weakness of bilateral lower extremities was improved so as to move against the gravity. But the next day, the limb weakness was aggravated as same as preoperative status due to mass effect of new hematoma. The second operation was performed to remove the hematoma and to control the bleeding focus. Several weeks later, the limb weakness was improved and he was able to walk. The literatures about spinal cavernous angioma are reviewed.

KEY WORDS : Epidural cavernous angioma · Cervicothoracic · Paraplegia · Magnetic resonance imaging.

Introduction

Cavernous hemangioma is a vascular malformation that may affect any part of the neuraxis. It has been reported involving the central nervous system simultaneously with other organs and may appear as sporadic or familial case.

Cavernous hemangiomas in a purely spinal epidural location are rare, comprising 1~2% of the cavernous hemangioma developed at the spine[6,11,15]. And spinal epidural cavernous hemangioma accounts for 4% of all spinal epidural tumor[10].

Spinal epidural cavernous hemangiomas present clinically as chronic or acute syndrome of spinal cord compression as well as local back pain or radiculopathy and must be considered in the differential diagnosis of other epidural diseases.

The aim of this article is to present a rare case of epidural cavernous hemangioma with unusual disease progression as sudden bilateral lower limb weakness in very young age, and to review the literature.

Case Report

A 23-month-old kid presented with sudden onset of both lower limb weakness. He had no perinatal problem and no abnormal developmental delay. Two days before visit, he got up at morning and was unable to move both lower limbs and to sit up by himself. On neurological examination, the limb power was grade II in both knee extension and flexion, and grade I in both ankle and toe movement. And the deep tendon reflexes in both legs were hyperreflexic.

Magnetic resonance imaging(MRI) revealed an epidural mass surrounding spinal cord associated with cord compression at the level of the C5 through T3. The mass was located mainly dorsal side of the cord, but at the level of the T1 to T3, the mass was surrounding the spinal cord. The mass was mainly isointense on T1 weighted images but lower part of the mass was hyperintense, and inhomogeneously hyperintense signal intensity on T2 weighted images and inhomogeneous enha-

• Received : June 15, 2006 • Accepted : July 6, 2006

• Address for reprints : Byung-Kyu Cho, M.D., Department of Neurosurgery, Seoul National University Hospital, 28 Yeongeon-dong, Jongno-gu, Seoul 110-744, Korea Tel : +82-2-2072-3639, Fax : +82-2-747-3648, E-mail : bkcho@snu.ac.kr

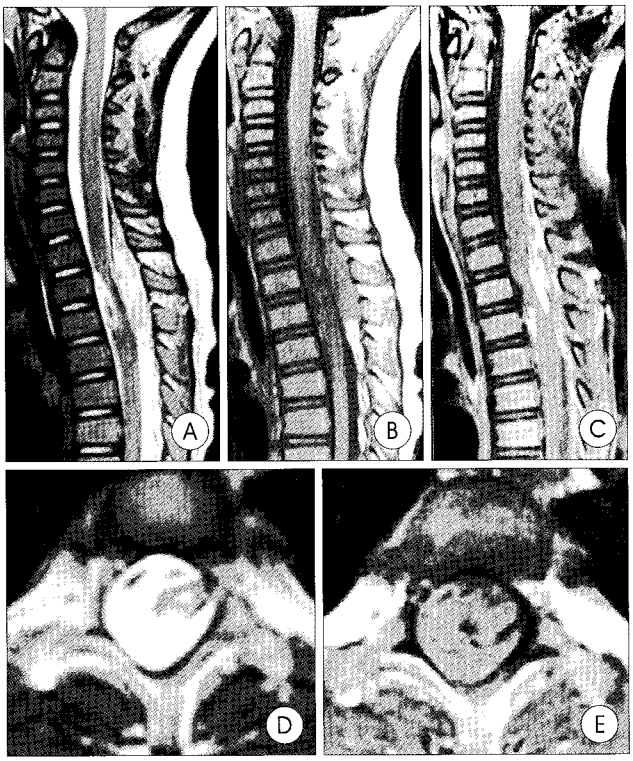

Fig. 1. Pre-operative magnetic resonance images of epidural cavernous angioma. A : Sagittal T2-weighted image shows heterogeneous high-signal intensity mass surrounding spinal cord at the level of the C5 through T3. B : Iso-signal intensity on T1-weighted sagittal image. C : Heterogeneous signal intensity on enhanced T1-weighted sagittal image. D : A high-signal intensity mass surrounding spinal cord displaced the cord left laterally on T2-weighted axial image. E : An iso-signal intensity epidural mass on T1-weighted axial image.

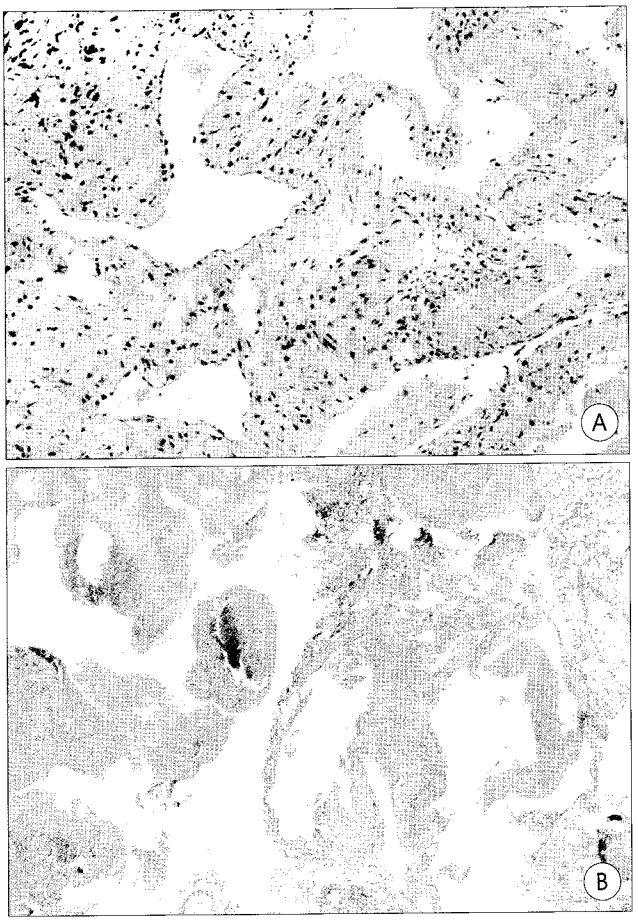

Fig. 2. Histologic finding of the epidural mass. The lesion is composed of multiple vascular channels, separated by intervening hyalinizing stroma (H&E, A : X200, B : X40).

ncement on enhanced-T1 weighted images (Fig. 1).

The lesion was approached through C6-T3 laminectomy. The lesion appeared as a dark red encapsulated mass. The lesion was moderately adherent to dural sac. Under the surgical microscope, it was possible to strip off the lesion from the dural sac. But the ventral portion of the mass was unable to remove completely, so the mass was removed subtotally. On neurologic examination after surgery, the lower extremity power was improved, the motor Grade III or IV. But, the next day, bilateral lower limb weakness was aggravated by the new epidural hematoma which was compressing the cord at the level of C7-T2 on follow-up MRI scan. So the second operation was performed with the same approach. By 2 weeks postoperatively, the limb power improved and the patient was able to walk. Histopathological diagnosis was cavernous hemangioma. Sections revealed closely opposed vascular channels lined with endothelium without any neural tissue in- between.

Discussion

A variety of names are used to describe cavernous hemangioma (cavernous angioma, hemangioma)[1]. Spinal epidural cavernous hemanigioma represent about 4% of all spinal epidural tumors. And spinal axis cavernous hamangioma, comprising vertebral, extradural, intradural extramedullary, and intramedullary lesions, constitutes 12% of all spinal vascular anormalies[6,10]. Solitary epidural cavernous malformations are exeeedingly rare compared with vertebral hemangiomas and represent 1~2% of all spinal cavernomas[3,6,11,15]. According to Antonio et al.[9], only 75 cases have been reported in English literature. And 4 cases in Korean literature were accessible to us[5,8,13]. Among them, this kid was the youngest. Clinical onset usually occurs during the 3rd to 6th decades of life and does not show any sex prevalence[1-4,14]. The segment most frequently affected is the thoracic one, followed by the lumbar and cervical one[1,2].

Like hamartomas, cavernous angiomas do not grow by mitotic activity. Growth of these lesions has been attributed to several mechanisms such as hemorrhage, dilation of the capillary bed, thrombotic-like processes, angioblastic proliferation or fusion of the vascular walls leading to the formation of real lacunae[7,14]. Clinical symptoms usually progress in a fairly insidious fashion and consist of sensory deficits, accompanied

Table 1. Differential diagnosis of an extradural lesion based on MR and CT findings

	T1-weighted MR	T2-wighted MR	Enhancement with Gd-DTPA	CT with bone window
Benign group				
Cavernous hamengioma	Iso- or hypointense	hyperintense	Homogeneous enhancement	Normal-sized neural foramen (usually)
Disc herniation	Iso- or hypointense	Hyperintense (rarely isointense)	None or peripheral enhancement	Normal-sized neural foramen (usually)
Schwannoma	Iso- or hypointense	Hyperintense	Heterogeneous enhancement with a rim	Enlarged neural foramen
Neurofibroma	Iso- or hypointense	Hyperintense	Homogeneous enhancement	Enlarged neural foramen
Angiolipoma	Hyperintense	Hyperintense	None or minimal enhancement (usually)	Normal-sized neural foramen
Osteochondroma	Hypointense	Hyperintense core with intermediate signal rim	None or minimal enhancement	Calcified mass within the foramen
Synovial cyst	Hypointense	Hyperintense	None or peripheral enhancement	Mass adjacent to facet joint; no bony change
Malignant group				
Metastasis Chordoma Ewing's sarcoma	Iso- or hypointense	Inhomogeneously hyperintense	Marked and homogeneous enhancement	Bony destruction

by radicular pain and/or paraparesis. But like this case, an acute or subacute onset can be occured and may be attributed to microhemorrhages or actual hematomas[1-4,7,15]. Furthermore, the resulting motor deficits may be severe.

Technological advances in diagnostic imaging techniques have made it possible to detect pathologies previously considered rare. MRI is the imaging modality of choice in detecting cavernous angioma. MRI image of the cavernous angioma is usually hypo- or isointense on T1 weighted images usually because of calcifications and iron deposits. On T2 weighted images, the cavernous angioma signal has a high intensity, just slightly less than that of cerebrospinal fluid. On enhanced T1 weighted images, the cavernous angioma signal shows generally homogeneous or slightly heterogeneous enhancement. Extraordinarily, because of the coexistence of hematoma in the acute and subacute phases, the portion of high signal intensity was seen on both T1 and T2 weighted images in this case. The hemosiderin rim was not obvious and this made the preoperative differential diagnosis more difficult.

Even though a histological diagnosis from radiological images cannot be predicted with accuracy for each and every case, there are several clues for differential diagnosis of spinal extradural solid lesions. The imaging characteristics of them are summarized in Table 1[10].

The correct preoperative diagnosis is important in order to plan and perform the best operation, also bacause intraoperative diagnosis of an epidural cavernous angioma is difficult, due to the limited exposure at the surgery and the epidural bleeding. The total resection is fundamentally recommended for the treatment of the cavernous angioma, but it may be incompletely stripped off and the complication of root injury may be provoked due to venous plexus bleeding, dense attachment or adhesion of the mass to the roots and its ventral location in part. For this incomplete resection of the mass, the radiotherapy may be helpful.[12] But because radiotheraphy is not without risk, we suggest that patients should instead be followed with frequent MRI assessments. A second-look surgery is clearly less risky than irradiation.

Conclusion

The authors report a case of spinal epidural cavernous hemangioma. It deserves an interest due to its rarity and can be also considered in the differential diagnosis of epidual spinal tumors even in young age.

References

1. Acciari N, Padovani R, Giuloni M, Gaist G : Surgical treatment of spinal cavernous angiomas. **J Neurosurg Sci 37** : 209-215, 1993
2. Goiwyn DH, Cardenas CA, Murtag FR, Balis GA, Klein JB : MRI of a cervical extradural cavernous hemangioma. **Neuroradiology 34** : 68-69, 1992
3. Graziani N, Bouillot P, Figarella-Branger D, Dufour H, Peragut J, Grisoli F : Cavernous angioma and arteriovenous malformation of the spinal epidural space. Report of 11 cases. **Neurosurgery 35** : 856-862, 1994
4. Hillman J, Bynke O : Solitary extradural cavernous hemangiomas in the spinal canal. **Surg Neurol 36** : 19-24, 1991
5. Hong SP, Cho DS, Kim MH, Shin KN : Spinal epidural cavernous hemangioma simulating a disc protrusion. A case report. **J Korean Neurosurg Soc 33** : 509-511, 2003
6. Jellinger K : Pathology of the spinal vascular tumours. In:Pia HW, Djindjian R (eds) : **Spinal angiomas : advances in diagnosis and therapy.** Springer : Wien New York 1978, pp 18-44
7. Lee JP, Wang AD, Wai YY, Ho YS : Spinal extradural cavernous hemangioma. **Surg Neurol 34** : 345-351, 1990
8. Park H, Park KS, Rhee DY : Two cases of epidural cavernous hemangioma in the thoracic spine. A case report. **J Korean Neurosurg Soc 16** : 921-927, 1987

9. Santoro A, Piccirilli M, Bristot R, di Norcia V, Salvati M, Delfini R : Extradural spinal cavernous angiomas : report of seven cases. **Neurosurg Rev 28 :** 313-319, 2005
10. Saringer W, Nobauer I, Haberler C, Ungersbock K : Extraforaminal, thoracic, epidural cavernous hemangioma : case report with analysis of magnetic resonance imaging characteristics and review of the literature. **Acta Neurochir (Wien) 143 :** 1293-1297, 2001
11. Shin JH, Lee HK, Rhim SC, Park SH, Choi CG, Suh DC : Spinal epidural cavernous hemangioma:MR findings. **J Comput Assist Tomogr 25 :** 257-261, 2001
12. Talacchi A, Spinnato S, Alessandrini F, Iuzzolino P, Bricolo A : Radiologic and surgical aspects of pure spinal epidural cavernous angiomas. Report on 5 cases and review of the literature. **Surg Neurol 52 :** 198-203, 1999
13. Yang YH, Kim HJ, Kang JK, Park YS : Spinal epidural cavernous hemangioma of the upper thoracic vertebrae. Case report. **J Korean Neurosurg Soc 32 :** 477-480, 2002
14. Zevgaridis D, Buttner A, Weis S, Hamburger C, Reulen HJ : Spinal epidural cavernous hemangiomas. **J Neurosurg 88 :** 903-908, 1998
15. Zevgaridis D, Medelele RJ, Hamburger C, Steiger HJ, Reulen HJ : Cavernous haemangiomas of the spinal cord. A review of 117 cases. **Acta Neurochir (Wien)** 141 : 237-245, 1999