www.jkns.or.kr 10.3340/jkns.2011.49.6.377 Print ISSN 2005-3711 On-line ISSN 1598-7876
J Korean Neurosurg Soc 49 : 377-380, 2011

Case Report

Spinal Intradural Extramedullary Cavernoma Presenting with Intracranial Superficial Hemosiderosis

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A case of intradural extramedullary cavernous angioma is presented with headache, dizziness, and bilateral sensorineural hearing loss caused by an intracranial superficial hemosiderosis. It was incidentally found in a patient with a 3-month history of sustained headache, dizziness and a 3-year history of hearing difficulty. The neurological examination was unremarkable in the lower extremity. MR images showed an intracranial superficial hemosiderosis mostly in the cerebellar region. Myelography and MR images of the thoracolumbar spine revealed an intradural extramedullary mass, which was pathologically proven to be a cavernous angioma. T12 total laminoplastic laminotomy and total tumor removal were performed without any neurologic deficits. The patient's symptoms, including headache and dizziness, have been absent for three years. Intradural extramedullary cavernous angioma can present with an intracranial superficial hemosiderosis as a result of chronic subarachnoid hemorrhage.

Key Words : Cavernous angioma · Spinal cord tumor · Superficial hemosiderosis.

INTRODUCTION

Intradural extramedullary cavernous angiomas (CAs), or cavernomas, of the spinal cord are rare entities. The most recent review of the literature showed twenty-eight cases of intradural extramedullary cavernomas[2,3,5,9,15], ten cases of which presented with spinal subarachnoid hemorrhage. The authors report a unique case of intradural extramedullary cavernoma of the cauda equina which results in an intracranial superficial hemosiderosis due to chronic recurrent subarachnoid hemorrhage.

CASE REPORT

A 55-year-old man was admitted to our hospital with a 3-month history of sustained headache and dizziness. Fifteen years prior he had experienced a severe headache that spontaneously resolved. Six years prior his eyesight became weaker. Three years prior he started experiencing decreased hearing in the left ear. The onset of these symptoms was gradual and the course was slowly progressive. There was no evidence of diabetes mellitus, hypercholesterolemia, or hypertension.

• Received : October 25, 2010 • Revised : December 9, 2010
• Accepted : May 30, 2011
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Neurologic examinations revealed no neurologic deficits except mild sensorineural hearing loss in the right ear and deafness in the left ear. Neurophysiological tests were carried out and vestibular evoked myogenic potential and a brain stem auditory evoked potential tests revealed no response in the left ear. A caloric test for vestibule-ocular reflex showed a normal response in both ears. The laboratory results were within normal range.

Brain magnetic resonance (MR) images disclosed severe cerebellar (hemispheric and vermian) atrophy with the hypointensity on gradient-echo T2-weighted images (Fig. 1). Cerebral angiography taken to clarify a hemorrhagic focus revealed no evidence of aneurysm or vascular malformation. In cerebrospinal fluid (CSF) analysis, the presence of red blood cells and xanthochromia was not detected. CSF glucose was normal. CSF protein was slightly elevated to 57 mg/dL. The opening pressure was normal at 110 mm H_2O. Myelography and MR images of the spine demonstrated an ovoid intradural extramedullary mass in the cauda equina on the level of the T12 vertebral body (Fig. 2). The lesion was slightly hyperintense on T2- and isointense on T1-weighted images without contrast enhancement compared to the instensity of the conus medullaris (Fig. 3).

A T12-L1 laminoplastic laminotomy was performed and the dura was opened. The dorsal arachnoid membrane was opaque and adhered to the rootlets. A 2.0×1.3×1.3 cm sized, dark-red blackberry-like subarachnoid tumor originated from a single rootlet and remained closely adherent to the rootlets in the left

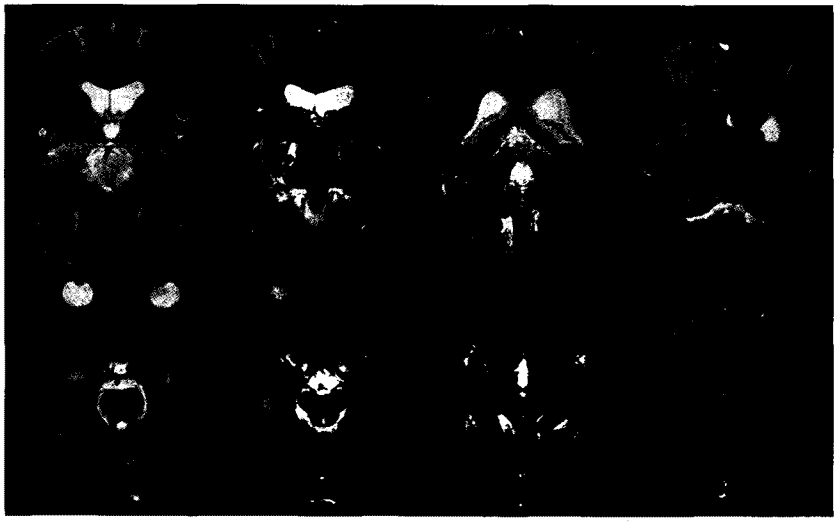

Fig. 1. Preoperative magnetic resonance gradient-echo T2-weighted image demonstrating the circumferential hypointensity due to hemosiderin deposition around the dorsal midbrain and cerebellar follia.

Fig. 2. Myelography showing the complete block of cerebrospinal fluid space at the level of the T12-L1 disc space.

Fig. 3. Sagittal images demonstrating a mass with an isosignal intensity on the T2-weighted, T1-weighted images and without enhancement on the gadolinium enhanced T1-weighted image.

side (Fig. 4). However, there was no evidence of marginal hemosiderosis around the mass. The lesion was totally excised after sacrificing a single rootlet on which the mass originated from a fine vessel. Histopathological examination showed the tumor to be a cavernous angioma. He noted complete resolution of his headache and dizziness immediately after the operation. Mild voiding difficulty and hypesthesia in the perineum and right medial thigh developed postoperatively. However, these symptoms disappeared after one month. The patient has been completely symptom free for three years.

DISCUSSION

Review of literature

CAs are benign hamartomatous vascular lesions. They consist of irregular sinusoidal vascular spaces lacking intervening neural or glial tissue and are lined by a single layer of endothelium. Most CAs originate from the abnormal development of periradicular vessels[5]. They are located mostly in the vertebral body. Only 3% of CAs are reported to be intradural[9]. Intradural extramedullary CAs are rarer than intramedullary ones. Until now, 29 cases of spinal intradural extramedullary CAs including our case have been reported in the literature[2,3,5,9,15]. According to the review data collected up to now (Table 1), the mean age at diagnosis is 47 (range 20-75). There is a male predominance (M : F=21 : 8) and the lesions are mostly located in the lumbar region (L2-5, 11 cases), followed by the thoracolumbar region (T12-L1, 8 cases), thoracic region (T1-11, 5 cases), and cervical region (5 cases). CAs originated mostly from rootlets (22 of 26 cases confirmed). Ten cases (34%) presented with significant subarachnoid hemorrhage. Two cases reported the development of a communicating hydrocephalus respectively with or without apparent bloody CSF. Low back pain and radiculopathy are the main symptoms associated with or without motor deficit or neurologic symptoms secondary to spinal cord compression. Although total excision was obtained except in one case, symptoms remained in seven cases (25%).

The uniqueness of our case

Superficial hemosiderosis results from hemosiderin deposition in the subpial layers of the brain and spinal cord[12]. The chronicity of bleeding is indispensible to the development of the hemosiderosis; it cannot occur after a single bleeding episode[1]. However, a source of bleeding of the central nervous system is found in more than half of all cases[6]. The underlying causes of superficial siderosis have included bilateral jugular vein thrombosis, primary angiosarcoma of the pineal gland, intraventricular cavernous

malformation, cerebral amyloid angiopathy, ependymoma of the cauda equina, and cervical root avulsion[7,8,11,13,17,19]. Procedural complications following ventriculoperitoneal shunt, hemispherectomy, pseudomeningocele formation after cervical decompressive surgery, and posterior fossa surgery have also been reported as iatrogenic causes[4,10,14,18].

The cerebellar cortex is also susceptible to hemosiderin deposition[1]. The special vulnerability of the eighth cranial nerve has been attributed to the long course of this nerve covered by central myelin in the subarachnoid space[4]. The classical manifestations of hemosiderosis include adult-onset slowly progressive gait ataxia and, less commonly, appendicular ataxia, ataxic dysarthria, and sensorineural hearing impairment[12].

Fig. 4. An intraoperative photograph showing a blackberry-like cavernous angioma originating from a fine vasculature on the rootlet.

Table 1. *Literature review of spinal intradural extramedullary cavernous angioma*

Author	Year	Age	Sex	Symptoms	Origin	Location	Extent of removal	Outcome
Roger et al.	1951	22	F	Sciatic and back pain, M deficit	ND	T11	T	Worse
Floris	1958	57	M	M deficit	ND	T12	T	ND
Hirsch et al.	1965	20	M	SAH, SM deficit, sphincter change	Root	L2-3	T	Remained
Pansini and Lo Re	1966	46	M	Sciatic and back pain, SM deficit, sphincter and erectile dysfunction	Root	L2	T	Remained
Ortner et al.	1973	22	M	SAH	ND	C4-7	T	Remained
Heimberger et al.	1982	24	M	SAH	Root	T2-3	T	Excellent
Ueda et al.	1987	28	M	SAH, pain	Root	L1-2	T	Excellent
Pagni et al.	1990	46	M	Back pain	Intraroot	T12-L1	T	Excellent
Ramos et al.	1990	67	F	Hydrocephalus, cognitive dysfunction, sphincter change, gait disturbance	Filum	L3	T	Excellent
Mastronardi et al.	1991	49	F	SM deficit	Root	T4	T	Excellent
Mori et al.	1991	65	M	SAH	Adherent to cord	T1	T	Excellent
Acciarri et al.	1992	54	F	SAH	Dura mater	C2-3	T	Excellent
Sharma et al.	1992	63	M	Back pain, SM deficit, sphincter change	Root/Cord	T12	T	Remained
Sharma et al.	1992	43	M	SAH	Root/Cord	T5	T	Excellent
Bruni et al.	1994	28	M	SAH	Root	L2	T	Excellent
Cervoni et al.	1995	26	F	SAH	Root	L1-2	T	Excellent
Cervoni et al.	1995	32	M	Pain	Root	L5	T	Remained
Makino et al.	1995	67	M	SAH, Hydrocephalus	Root	L2	T	Excellent
Rao et al.	1997	60	M	SM deficit	Root	L1-3	T	Excellent
Rao et al.	1997	35	F	SM deficit	Adherent to cord	T12	ST	Remained
Duke et al.	1998	49	F	Sciatic and back pain, S deficit	Root	L4	T	Excellent
Nozaki et al.	2003	51	M	SM deficit	Root/Dentate ligament	C5-6	T	Excellent
Falavigna et al.	2005	44	F	Leg numbness, sphincter change	Intraroot	L4	T	Excellent
Rachinger et al.	2006	56	M	Shoulder pain	Root	C7	T	Excellent
Caroli et al.	2007	71	M	Sciatic and back pain, S deficit	Intraroot	L3	T	Excellent
Er et al.	2007	67	M	Sciatic and back pain, SM deficit, sphincter change	Root	T12-L2	T	Excellent
Cecchi et al.	2007	75	F	Paresthesia in both legs	Root	L3-4	T	Excellent
Kivelev et al.	2008	44	M	Brown-Sequard syndrome	Root	C5-6	T	Excellent
Jin et al.	2010	55	M	SNHL (hemosiderosis), headache, dizziness	Root	T12-L1	T	Excellent

M : male, F : female, ND : not described, SM : sensorimotor, SAH : subarachnoid hemorrhage, SNHL : sensorineural hearing loss, T : total, ST : subtotal

To our knowledge, this is the first case report of superficial hemosiderosis related to a cavernous hemangioma of the cauda equina. In our patient, the hemosiderin deposition was a consequence of recurrent and persistent bleeding into the subarachnoid space by the spinal intradural extramedullary CA. Er et al.[5] suggested subarachnoid hemorrhage may present as the initial symptom of intradural extramedullary cavernous angioma. Detachment of the adherent mass from the nerve roots may result from the dynamic vertebral canal which is coupled to its restricted mobility. Although there was no evidence of active bleeding at diagnosis in the present case, superficial hemosiderosis was evident in the cerebellar folia. The sensorineural hearing difficulty was due to such pathology. However, there was no evidence of recent bleeding at diagnosis. Therefore, it is possible that symptoms associated with hemosiderosis can be initial presentations of hidden intraspinal CAs in patients with subclinical symptoms in spite of recurrent subarachnoid hemorrhage.

It is especially interesting for a patient to recover from headache immediately after tumor removal. Such clinical finding seems to indicate the cause and effect relationship between the tumor and headache. That also means that the headache was not related to hemosiderosis. One of two patients with hydrocephalus in the literature presented with normal pressure hydrocephalus without active subarachnoid hemorrhage at diagonsis[16]. Ramos et al.[16] explained diverse causative factors such as mechanical obstruction in CSF flow due to the tumor, hyperviscosity secondary to increased CSF albumin, slowed CSF flow, and partitioning of CSF absorption sites from an inflammatory reaction. Although there was no radiologic finding indicating the development of hydrocephalus and opening pressure was normal, the authors believe that the pathogenesis of headache and dizziness may be related to a dynamic CSF pathway blockade which created a pressure gradient between the proximal and the distal subarachnoid space and provoked an insufficient buffering of intracranial pressure. The normal opening pressure at the L4-5 level may be attributed to a pressure gradient. The immediate resolution of such symptoms after the restoration of the CSF pathway supports that idea instead of the relationship to hemosiderosis.

Intermittent headache by dynamic CSF blockade due to tumor and sustained sensorineural hearing loss caused by superficial hemosiderosis as a result of recurrent subarachnoid hemorrhage can be possible manifestations in patients with intradural extramedullary cavernous angiomas. Therefore, spine MR images should be considered in patients with superficial hemosiderosis when a bleeding focus has not been detected intracranially.

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