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$_2$O. 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 intensity 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...

**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.

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.

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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...
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. They are located mostly in the vertebral body. Only 3% of CAs are reported to be intradural. 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. 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. The chronicity of bleeding is indispensable to the development of the hemosiderosis; it cannot occur after a single bleeding episode. However, a source of bleeding of the central nervous system is found in more than half of all cases. 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. Procedural complications following ventriculoperitoneal shunt, hemispherectomy, pseudomeningocele formation after cervical decompressive surgery, and posterior fossa surgery have also been reported as iatrogenic causes.

The cerebellar cortex is also susceptible to hemosiderin deposition. 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. The classical manifestations of hemosiderosis include adult-onset slowly progressive gait ataxia and, less commonly, appendicular ataxia, ataxic dyshartria, and sensorineural hearing impairment.

**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    |
| Ortnet 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                  | Intra-root   | T12-L1   | T                 | Excellent   |
| Ramos et al.           | 1990 | 67  | F   | Hydrocephalus, cognitive dysfunction, sphincter change, gait disturbance | Filum        | L3       | T                 | Excellent   |
| Mastromardi 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 | Intra-root   | 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 | Intra-root   | 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. 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 diagnosis. Ramos et al. 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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