Magnetic Resonance Imaging of Unusual Intramedullary Spinal Cord Lesions

Nadir Intrameduller Spinal Kord Lezyonlarında Manyetik Rezonans Görüntüleme

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Abstract
Magnetic resonance imaging is the current imaging modality of choice for evaluating patients presenting with myelopathic symptoms in the search for spinal cord lesions. Early diagnosis plays an important role in the management of these lesions and can affect the prognosis and final outcome of the patient. In this review article, the clinical presentation and magnetic resonance signal characteristics of unusual intramedullary spinal cord lesions, including infarction, cavernous malformation (CM), arteriovenous malformation (AVM), tuberculoma, brucella abscess, hemangioblastoma, dermoid, epidermoid tumors, and metastases, are presented.

Keywords
MR Imaging; Spinal Cord; Intramedullary Lesions


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Introduction
Spinal cord lesions are rare; magnetic resonance imaging should be performed as soon as possible and as the first technique whenever an intrinsic spinal cord lesion is suspected clinically. In this pictorial essay, unusual intramedullary spinal cord lesions, including vascular lesions of the spinal cord (such as infarction, cavernous malformation (CM), arteriovenous malformation (AVM), spinal cord abscess (including tuberculosis and brucella abscess) and rare intramedullary neoplasms (including hemangioblastoma, dermoid, epidermoid tumors, and metastases) are presented. The clinical presentation and magnetic resonance signal characteristics of these unusual intramedullary lesions are discussed.

Vascular Diseases of the Spinal Cord
Vascular diseases affecting the spinal cord, while a relatively rare occurrence, compared with cerebrovascular events, can cause substantial neurological morbidity. They include various pathologies, including structural issues (e.g., infarction, cavernous malformation (CM), arteriovenous malformation (AVM)).

Spinal Cord Infarction
The key to distinguishing spinal cord infarction from other entities that can have a similar appearance on imaging is the clinical history. Spinal cord infarction is characterized clinically by sudden motor and sensory loss below the level of the spinal cord injury [1]. Underlying causes include aortic disease and surgery, atherosclerosis, degenerative spinal disease, systemic hypotension, vertebral artery dissection, coagulopathy, trauma, and cocaine abuse. However, the cause has been reported to be idiopathic in 28-74% of cases [2]. Infarction occurs most commonly in the thoracic and thoracolumbar cord, which has a tenuous blood supply. Spinal cord infarction typically manifests in MRI as focal cord swelling, including intramedullary hyperintensity on T2-weighted images (Fig. 1). Diffusion-weighted imaging demonstrating focal restricted diffusion has become increasingly sensitive for the diagnosis of spinal cord infarcts, but its sensitivity is not as high as in the brain. Infarction may show minimal enhancement (typically after 5 days). Contrast enhancement may persist for up to 3 weeks after onset. Mild cord expansion is often present. However, as described in several studies, conventional MRI may appear normal in some patients, especially in the acute phase [1-4].

Cavernous Malformation
Cavernous malformation (CM) is also known as cavernous angioma, cavernous hemangioma, and cavernoma. CMs are angiographically occult vascular malformations, defined by abnormal, enlarged vascular channels but without interposing neural or glial tissue. Intracerebral CMs are more common than spinal lesions; coexisting intracranial CMs can be found in a quarter of cases and even more frequently in familial cases [3]. The clinical presentation is variable, and patients may receive a diagnosis of an asymptomatic intramedullary CM. The acute presentation is likely due to hemorrhage within the vascular spaces or into the spinal cord parenchyma [1].

The diagnosis of spinal CM requires MRI. CM typically demonstrates a speckled "popcorn" heterogeneous signal intensity on both T1- and T2-weighted images due to blood products in various stages of evolution. The lesion may demonstrate a peripheral rim of hemosiderin of low signal intensity on T2-weighted images. Gradient echo sequences may demonstrate the "blooming" susceptibility artifact from the presence of blood products (Fig. 2) [1,3,4]. CT may also be of value, be-

Figure 1. Spinal cord infarction. A 55-year-old female with recent abdominal aortic aneurysm repair presented with paralysis and paresthesia in both lower extremities. MR image obtained 2 days after the onset of symptoms. Sagittal T2-weighted (A), image demonstrates diffuse central hyperintensity (arrows) with cord expansion between the Th11 and L1 spinal levels. Axial T2-weighted (B), hyperintense signal with involvement of gray matter and adjacent central white matter (arrow).

Figure 2. Cavernous malformation. A 61-year-old female with an incidentally revealed spinal cord cavernous malformation (CM). Sagittal T1-weighted (A) and T2-weighted (B) spinal MR images show a focal area of hyperintensity with a hypointense rim on the right side of the cervical cord (arrow). (C) The lesion "blooming" artifact on a sagittal gradient echo image (arrow).
resent upwards of 70% of all spinal AVMs [1]. Both sdAVFs and AVMs are predominantly thoracolumbar, but may develop at any level of the cord, including the filum terminale. They cause symptoms primarily from venous hypertension, which then produces cord edema and, eventually, infarction. Spinal MRI and angiography are essential in establishing a diagnosis of spinal AVM. Spinal MRI has excellent sensitivity and specificity for the detection of spinal AVMs and sdAVF, as long as it includes all necessary sequences and is interpreted by experienced neuroradiologists [3]. MRI of the entire spine should be performed, be it in the setting of a progressive myelopathy or spinal subarachnoid hemorrhage, because neurological signs and symptoms do not correspond well with the level of the vascular lesion. MRI characteristics of the myelopathy associated with sdAVF include longitudinally extensive T2 hyperintensity within the central aspect of the spinal cord, involving the conus medullaris in as many as 95% of patients. Spinal cord enlargement and dilated perimedullary veins may also be seen, most prominently on sagittal T2 sequences. The cord becomes swollen and may demonstrate patchy or diffuse enhancement as a sign of chronic venous congestion [4] (Fig. 3).

Spinal Cord Abscess
Intramedullary abscess of the spinal cord is a rare infection of the central nervous system. It is associated with high morbidity and mortality. Early recognition and treatment are important [5]. Multiple organisms have been shown to cause intramedullary abscess; we discuss tuberculoma and brucella abscess.

Tuberculoma
The most common form of spinal tuberculosis is meningitis, but involvement of spinal cord in the form of intramedullary tuberculoma is rare and the concurrent occurrence of cranial and intramedullary tuberculomas is extremely rare [6]. Central nervous system tuberculosis occurs because of the hematogenous spread of the bacilli from a distant focus. Additionally, in spinal tuberculosis, the tuberculous bacilli may gain entry through the subarachnoid space or central canal and produce a local inflammatory response, which can evolve into an intramedullary granulomatous lesion [6,7]. The clinical presentation of intramedullary tuberculoma is progressive myelopathy, with or without sphincter disturbances [6,8]. The MRI findings in cases of spinal intramedullary tuberculoma can vary during the different phases of the tuberculoma. In the early phase, there are severe infective reactions and variable degrees of edema around the lesion. During this phase, tuberculoma appears isointense on both T1WI and T2WI and is homogenously enhanced. Subsequently the tuberculoma capsule becomes richer in collagen. As a result, T1WI shows equal signal intensity and T2WI shows equal or low signal intensity. After enhanced scanning, there is rim enhancement and a low signal in the central region (Fig. 4). The center of the tuberculoma...
becomes hyperintense on T2WI with the development of caseation. The solid parts of granulomas may appear as hypo- to hyperintense on T2WI with the development of caseation; also, T2WI typically shows a “target sign” [7,8].

Brucella Abscess
Brucellosis is an infectious disease caused by a Gram-negative intracellular coccobacillus. Brucella reaches the CNS via the hematogeneous route or by direct extension from spondylitis. Nervous system complications of neurobrucellosis include meningitis, encephalitis, brain abscess, epidural abscess, demyelination syndromes, and meningovascular syndromes. Meningitis has been reported as the most frequent presentation, occurring in ~50% of cases, but abscess and granuloma development generally have rarely been reported [9,10]. Neurobrucellosis can be diagnosed by raised brucella titers in the CSF, which were also detected in the present case. The lesion exhibits a hypointense signal on T1-weighted sequences and a hyperintense signal on T2-weighted sequences with associated spinal cord edema (Fig. 5). Intramedullary abscesses may show restricted diffusion on diffusion-weighted images [5].

Rare Intramedullary Neoplasms
We discuss hemangioblastomas, dermoid and epidermoid tumors, and metastases.

Hemangioblastoma
Hemangioblastomas are benign WHO grade I tumors. Hemangioblastomas of the spinal cord tend to be solitary (80%), with multiple lesions being suggestive of VHL syndrome (autosomal-dominant cerebelloretinal hemangioblastomatosis, renal cell carcinoma, multiple renal and pancreatic cysts, pancreatic cyst-adenocarcinoma, and epididymal cysts) [1]. These neoplasms are slow-growing and may present with sensory changes, pain, and motor dysfunction [11]. On MRI, hemangioblastomas are well-circumscribed nodular masses with variable T1 intensity (typically isointense), prominent enhancement with gadolinium, and hyperintensity on T2-weighted imaging. Flow voids, adjacent cysts, and hemorrhage are also common findings. The signal within the cyst may vary depending on the protein content within the cyst fluid. A syrinx may be present, and there may also be long segment cord edema without a syrinx [11,12]. Some cases may have the classic “cystic mass with an enhancing mural nodule” appearance characteristic of cerebellar hemangioblastoma (Fig. 6) [1].

Dermoid Tumor
Dermoid tumors or dermoid cysts are rare, congenital, benign, slow-growing tumors composed of more than one of the three primitive germ cell layers that produce skin and its appendages (hair follicles, sweat gland, and sebaceous glands). Dermoid cysts can be associated with dermal sinus (20%), vertebral abnormalities, and closed dysraphism [1]. Patients may present with slowly progressive compressive radiculopathy and myelopathy or cauda equina syndrome. These tumors may often become acutely symptomatic after rupture or infection [1,13]. On MR imaging, the signal intensity characteristics are variable, depending on the cystic contents, with the two major components being fluid and lipid. The lipid components appear hyperintense on T1-weighted MR images and exhibit low signal in-tensities on gadolinium-enhanced fat-suppressed...
T1-weighted MR images (Fig. 7). The lesion can also appear hypointense on T1-weighted MR images because of increased water content. On T2 weighted MR images, the fluid components are hyperintense. The soft-tissue component usually enhances after administration of intravenous contrast [1,13].

**Epidermoid Cysts**

Epidermoid cysts are benign tumors originating from ectoderm remnants. Intramedullary epidermoid cysts are very rare entities. They can be congenital, and are frequently associated with other spinal malformations, such as spina bifida [14]. Patients may present with non-specific presentations and symptoms, such as numbness, weakness, spasticity, and paraparesis of the lower extremities [15]. On MRI, where the absence of peritumoral edema and sharp boundaries between the lesion and surrounding parenchyma define the diagnosis, there is minimal peripheral enhancement with gadolinium, and an inhomogeneous signal is usually observed on both T1 and T2WI [14,15]. At MR imaging, epidermoid tumors can mimic cystic lesions with fluid content, such as arachnoid cysts. DWI can help in obtaining a correct diagnosis. Arachnoid cysts exhibit elevated diffusion with low signal on trace DWI and high ADC values, whereas epidermoid cysts show high signals on DWI (Fig. 8). ADC measured in the tumor was lower than that reported in intracranial epidermoids [14].

**Metastasis**

Intramedullary spinal cord metastases (ISCM) are rare. Although lung and breast cancers are the most common tumor sites in ISCM, melanoma, renal, colorectal, and lymphoma have also been reported [1,12]. The prognosis of patients with ISCM is very poor and nearly all patients present with motor weakness and pain, as well as bowel and bladder dysfunction (60%) and parasthesias (50%) [1]. At MR imaging, metastatic intramedullary lesions often show cord expansion due to associated edema, which is usually out of proportion to a seemingly focal small cord lesion, usually extending over several vertebral seg-

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**Figure 7.** Intramedullary dermoid tumor. A 3-year-old female patient with a previous history of occult spinal dysraphism. Sagittal MRI of spine: T2-weighted (A), T1-weighted (B) and gadolinium-enhanced fat-suppressed T1-weighted images (C). There is a well-defined intramedullary cystic mass, expanding the dorsal spinal cord. The dorsal component (arrows) is hyperintense on T1-weighted and T2-weighted images and shows suppression on gadolinium-enhanced fat-suppressed T1-weighted MR image, consistent with fat.

**Figure 8.** Intramedullary epidermoid cysts. A 3-year-old male patient who presented with bilateral lower extremity weakness for 4 days. MRI of the thoracolumbar spine shows an intramedullary mass at the T8-9 level. On sagittal T2-weighted image (A), the intramedullary lesion appeared hyperintense and well-defined (arrow). Sagittal T1-weighted image (B) lesion hypointense (arrow). On sagittal DWI (C), a marked high signal with low apparent diffusion coefficient value (D) is demonstrated. Postcontrast images may show either a ring-enhancing or a homogenously enhancing lesion (Fig. 9). The presence of hemorrhage may produce heterogeneous enhancement [1,12].

**Figure 9.** Intramedullary metastasis (primary, lung). A 45-year-old female with lung cancer with a known history of metastatic disease presents with bilateral lower and upper extremity paresthesia and weakness. On sagittal T2-weighted image (A), the intramedullary lesion appears isointense (arrow), there is a diffuse increased T2 signal, consistent with edema with enlargement of the cord (arrow head). Postcontrast sagittal (B) images show a lesion (arrow) of homogeneous enhancement in the cervical cord.
Conclusion
We have briefly reviewed clinical presentation and magnetic resonance signal characteristics of unusual intramedullary lesions, including infarction, CM, AVM, tuberculoma, brucella abscess, hemangioblastoma, dermoid, epidermoid tumors, and metastases. When spinal cord diseases are suspected, MRI should constitute the first diagnostic modality to identify the lesion and rule out potential differential diagnoses.

Competing interests
The authors declare that they have no competing interests.

References

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