Magnetic Resonance Imaging of Unusual Intramedullary Spinal Cord Lesions

Nadir Intrameduller Spinal Kord Lezyonlarında Manyetik Rezonans Görüntüleme

Spinal Kord Lezyonlarında Manyetik Rezonans Görüntüleme / Magnetic Resonance Imaging of Spinal Cord Lesions

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Özet

Manyetik rezonans görüntüleme, miyelopatik semptomlar ile başvuran spinal kord lezyonu araştırılan hastaların değerlendirilmesi için tercih edilen geçerli görüntüleme yöntemidir. Erken tanı, bu lezyonların tedavisinde önemli bir rol oynar, prognozu ve hastanın sonucunu etkileyebilir. Bu derlemede infarkt, kavernöz malformasyon (CM), arteriovenöz malformasyon (AVM), tüberküloma, brusella apsesi, hemangioblastoma, dermoid, epidermoid tümörler, ve metastazı içeren nadir intramedüller spinal kord lezyonları klinik bulgular ve manyetik rezonans sinyal özellikleri sunulmuştur.

Anahtar Kelimeler

MR Görüntüleme; Spinal Kord; İntrameduller Lezyon

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 tuberculoma 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, coag¬ulopathy, 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].

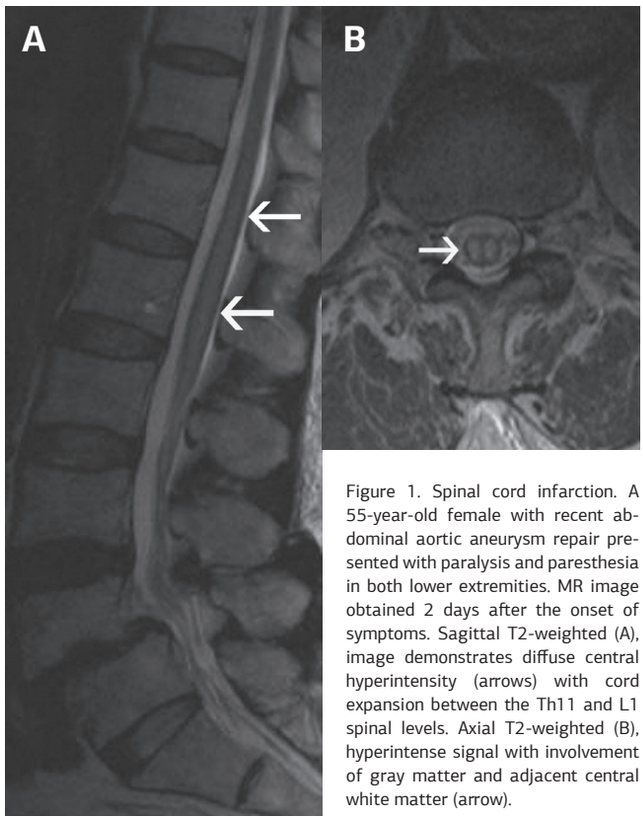


Figure 1. Spinal cord infarction. A 55-year-old female with recent abdominal aortic aneurysm repair presented with paralysis and paresthesia in both lower extremities. MR image obtained 2 days after the onset of symptoms. Sagittal T2-weighted (A), image demonstrates diffuse central hyperintensity (arrows) 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).

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, because some cavernomas demonstrate extensive calcification. In acutely hemorrhaging cavernomas, however, spinal angiography should be performed to rule out small glomerular AVM [1].

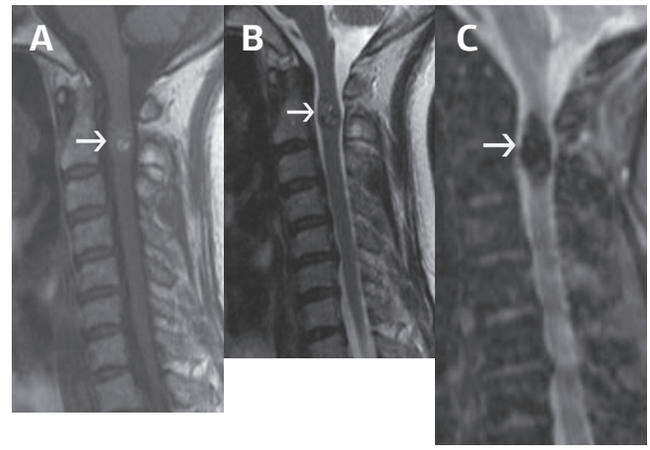


Figure 2. Cavernous malformation. A 61-year-old female with an incidentally revealed spinal cord cavernous malformation (CM). Sagittal T1- weighted (A) and T2- weighted (B) spinal MR images show a focal area of hyperintensity with a hypointense rim on the right side of the cervical cord (arrow). (C) The lesion "blooming" artifact on a sagittal gradient echo image (arrow).

Spinal AVMs

Spinal cord AVMs are congenital lesions resulting from a defect in early vascular embryogenesis. Spinal AVMs are a rare clinical phenomenon, although precise estimates of the incidence are lacking for all but a specific subset called "spinal dural AV fistulae" (sdAVF), which has been reported to affect 5 10 patients per million population annually [3] . Spinal dural AV fistulae are the most frequent vascular malformations of the spine and rep-

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).



Figure 3. Spinal dural AV fistula: A 46-year-old male presented with bilateral lower extremity weakness for 2 months. Sagittal T2- weighted (A) spinal MR images, intramedullary high-intensity area (arrow) and dilated perimedullary vessels (open arrow), which are present as a flow void in these images. The combination of pathologically dilated vessels and edema of the cord is pathognomonic for spinal dural AV fistulae. Gadolinium-enhanced fat-suppressed T1 -weighted image (B) the cord is swollen, demonstrating patchy enhancement (arrow).

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



Figure 4. Co-occurrence of intracerebral tuberculoma with spinal cord tuberculoma. A 31-year-old female with a history of tuberculous meningitis with weakness right lower limb for 2 days. Sagittal T2- weighted (A) MR image showing an intramedullary lesion with extensive surrounding edema. Enhanced sagittal T1- weighted image (B) shows obvious ring enhancement of the intramedullary lesion (arrow).

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, demyelization 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].



Figure 5. Intramedullary Brucella abscess. A 15-year-old male with a history of brucellosis presented with progressive upper and lower extremity pins and needles symptoms. Pretreatment: Sagittal T2-weighted MR image (A) showing intramedullary isointense lesion with surrounding edema. Enhanced sagittal and axial T1-weighted (B,C) showed obvious homogeneous enhancement of the intramedullary lesion. The lesion is hyperintense on axial DWI (D). Restricted diffusion is seen on the ADC map (E).

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 cystadenocarcinoma, 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).

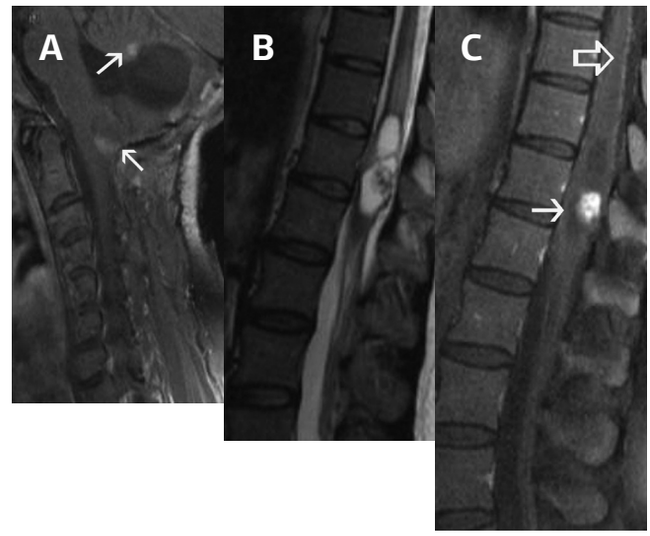


Figure 6. A 29-year-old female with multiple hemangioblastoma lesions in the spinal cord and cerebellum in von Hippel-Lindau (VHL) syndrome. Sagittal postcontrast T1- weighted images (A) show enhanced nodule posterior aspects of the cord and cerebellum (arrows). Sagittal T2-weighted (B) consistent with edema and a cystic mass. Sagittal postcontrast T1-weighted images (C) enhanced nodule (arrow) and open arrow show a hyperintense linear structure consistent with a vessel posterior to the cord.

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

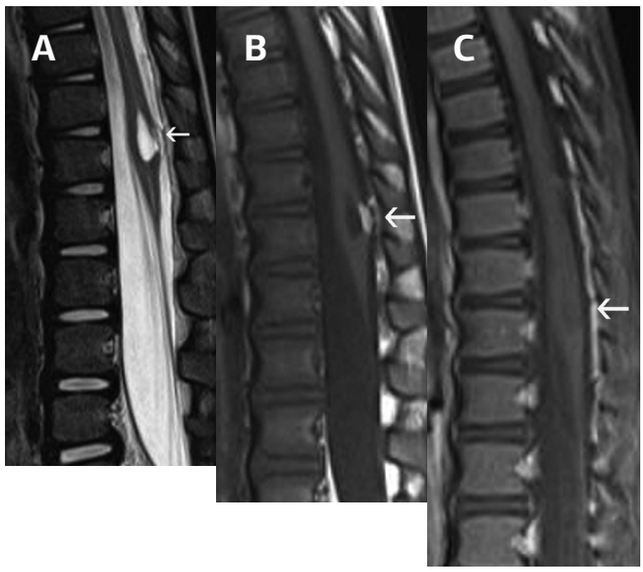


Figure 7. Intramedullary dermoid tumor. A 3-year-old female patient with previous history of occult spinal dyraphism. Sagittal MRI of spine: T2-weighted (A), T1-weighted (B) and gadolinium-enhanced fat-suppressed T1-weighted images (C). There is a well-defined intramedullary cystic mass, expanding the 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.

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 confine 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 segments. 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].

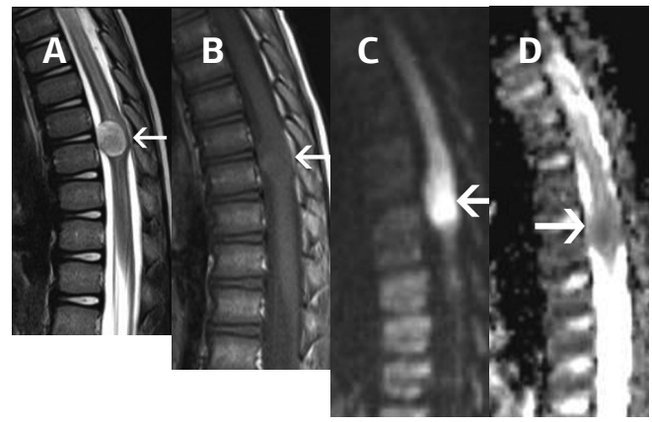


Figure 8. Intramedullary epidermoid cysts. A 3-year-old male patient who presented with bilateral lower extremity weakness for 4 days. MRI of the thoracolumbar spine shows an intramedullary mass at the T8-9 level. On sagittal T2-weighted image (A), the intramedullary lesion appeared hyperintense and 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.

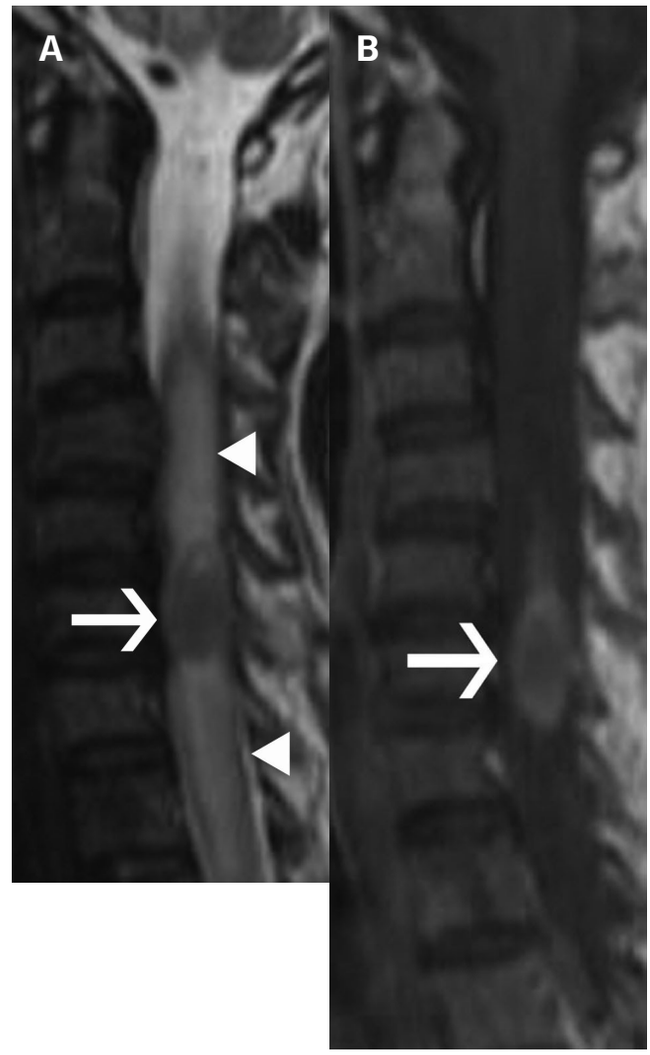


Figure 9. Intramedullary metastasis (primary, lung). A 45-year-old female with lung cancer with a known history of metastatic disease presents with bilateral lower and upper extremity paresthesia and weakness. On sagittal T2-weighted image (A), the intramedullary lesion appears isointense (arrow), there 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

1. Do-Dai DD, Brooks MK, Goldkamp A, Erbay S, Bhadelia RA. Magnetic resonance imaging of intramedullary spinal cord lesions: a pictorial review. Curr Probl Diagn Radiol 2010;39(4):160-85.
2. Alblas CL, Bouvy WH, Lycklama À Nijeholt GJ, Boiten J. Acute spinal-cord ischemia: evolution of MRI findings. J Clin Neurol 2012; 8(3):218-23.
3. Rubin MN, Rabinstein AA. Vascular diseases of the spinal cord. Neurol Clin 2013;31(1):153-81.
4. Krings T, Lasjaunias PL, Hans FJ, Mull M, Nijenhuis RJ, Alvarez H, et al. Imaging in spinal vascular disease. Neuroimaging Clin N Am 2007;17(1):57-72.
5. Roh JE, Lee SY, Cha SH, Cho BS, Jeon MH, Kang MH. Sequential magnetic resonance imaging finding of intramedullary spinal cord abscess including diffusion weighted image: a case report. Korean J Radiol 2011;12(2):241-46.
6. Chitre PS, Tullu MS, Sawant HV, Ghildiyal RG. Co-occurrence of intracerebral tuberculoma with lumbar intramedullary tuberculoma. J Child Neurol 2009;24(5):606-09.
7. Lu M. Imaging diagnosis of spinal intramedullary tuberculoma: case reports and literature review. J Spinal Cord Med 2010;33(2):159-62.
8. Torii H, Takahashi T, Shimizu H, Watanabe M, Tominaga T. Intramedullary spinal tuberculoma. Neurol Med Chir 2004;44(5):266-68.
9. Nas K, Tasdemir N, Cakmak E, Kemaloglu MS, Bukte Y, Geyik MF. Cervical intramedullary granuloma of Brucella: a case report and review of the literature. Eur Spine J 2007;16(3):255-59.
10. Bingöl A, Yücemen N, Meço O. Medically treated intraspinal "Brucella" granuloma. Surg Neurol 1999;52(6):570-76.
11. Smith AB, Soderlund KA, Rushing EJ, Smirniotopolous JG. Radiologic-pathologic correlation of pediatric and adolescent spinal neoplasms: Part 1, Intramedullary spinal neoplasms. AJR Am J Roentgenol 2012;198(1):34-43.
12. Mechtler LL, Nandigam K. Spinal cord tumors: new views and future directions. Neurol Clin 2013;31(1):241-68.
13. Altay H, Kitiş O, Calli C, Yunten N. A spinal dermoid tumor that ruptured into the subarachnoidal space and syrinx cavity. Diagn Interv Radiol 2006;12(4):171-73.
14. Thurnher MM. Diffusion-weighted MR imaging (DWI) in two intradural spinal epidermoid cysts. Neuroradiology 2012;54(11):1235-36.
15. Fereydoonian NA, Bakhti S, Fereshtehnejad SM, Tabibkhooei AR. Intramedullary thoracic spine epidermoid cyst with myelopathic presentations: a report of a rare case. Clin Neurol Neurosurg 2013;115 (6):841-43.

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