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Original Article

Intramedullary Spinal Lesions Involving the Conus Medullaris: MR Imaging Features for Differential Diagnosis

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Purpose : Intramedullary spinal lesions in the conus medullaris (CM), including tumors and vascular lesion, are rarely reported. We reported various MR features of intramedullary spinal cord lesions involving the CM including ependymoma, hemangioblastomas, dermoid cyst, ventriculus terminalis and spinal AVF and tried to discuss them for differential diagnosis.

Materials and Methods: Six patients (male: female = 4:2, mean age = 44.3 year old) were enrolled from the clinical database of our institute from 2004 to 2010 and their radiological images and clinical symptoms were reviewed retrospectively. All patients had taken initial and postoperative MRI with contrast enhancement using gadopentate dimeglumine (Gd-DTPA). These images were analyzed by tumor size, location, signal intensity relative to the spinal cord, vascular flow voids, syrinx or cyst, edema and enhancement pattern.

Results: Contrast enhancement was seen in all intramedullary masses. An eccentric enhancing nodule was noted in two hemangioblastomas and unusual peripheral rim enhancement with septation was seen in ventriculus terminalis. Patchy enhancement of the CM was observed in spinal arteriovenous fistula (AVF). Extensive cord edema adjacent to the intramedullary lesions was seen in four cases and syrinx was noted in three cases. Vascular signal voids were found in two hemangioblastomas and one spinal AVF.

Conclusion: In evaluation of intramedullary spinal lesions in the CM, it is necessary to consider these unusual MR findings and discriminate various pathologies with prudence and caution.

Index words : Conus medullaris · Intramedullary lesion · Spinal arteriovenous fistula · Ventriculus terminalis Hemangioblastoma · Dermoid cyst · Ependymoma

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INTRODUCTION

Intramedullary spinal lesions in the conus medullaris (CM), including tumors, infection and vascular lesion, are rarely reported. Intramedullary spinal cord tumors account for 1% of all central nervous system tumors (1) and some infectious diseases in the CM were reported as case reports (2–10). Moreover some authors described syringomyelia of the CM due to

spinal arteriovenous malformation (11). In a previous literature, 26.8% of 447 ependymomas were commonly located in the CM (12). Although they had a nonspecific radiologic appearance, Kim et al. (13) suggested some diagnostic clues of ependymoma were central location, diffuse enhancement, syringomyelia, hemorrhage and cap sign using MR imaging.

MR imaging is currently used as an imaging modality of choice in defining intramedullary spinal cord lesions (14, 15). MR imaging of the spinal cord tumor includes evaluation of tumor characteristics, the extent of cord involvement, enhancement pattern (14, 16).

Therefore we reported various MR features of intramedullary spinal cord involving the CM in six patients and tried to discuss them for differential diagnosis.

MATERIALS AND METHODS

This retrospective study was approved by our institutional review board and the requirement for the informed consent was waived. Six patients (male: female = 4:2, mean age = 44.3 year old) were enrolled from the clinical database of our institute from 2004 to 2010 and their radiological images and clinical symptoms were reviewed retrospectively. Intramedul-lary CM tumor was defined as the primary spinal cord tumor involved only at CM. Pathology was confirmed in five patients after total surgical resection (Table 1).

All patients had taken initial and postoperative MRI with contrast enhancement using gadopentate dimeglumine (Gd-DTPA). Lumbar-sacral MR imaging was performed using 1.5-T system (Vision 1.5T and Avanto 1.5T; Simens medical systems, Erlangen, Germany). Axial and sagittal T1, T2-weighted fast spin-echo sequence and contrast enhanced T1 -weighted images were taken in 1.5-T system (T1WI: TR/TE=560/9.8 msec; number of sections, 17; section thickness, 3 mm; field of view, 35 cm; matrix, 314 × 448; number of signals acquired, two; echo train length, 3; and voxel resolution, 1.1 × 0.8 × 3.0 mm, T2WI: TR/TE=3760/100 msec; number of sections, 17; section thickness, 3 mm; field of view, 35 cm; matrix, 338 × 512; number of signals acquired, four; echo train length, 3; and voxel resolution, ; 1.0 × 0.7 × 3.0 mm). These images were reviewed independently by two experienced neuroradiologists (S.J.A, T. S.C) and analyzed by the following parameters; tumor size, location, signal intensity relative to the spinal cord, vascular flow voids, syrinx or cyst, edema and enhancement pattern.

Table 1. Summary of Intramedullary Spinal Lesions in the Conus Medullaris

No	Sex	Age	Pathology	Symptom	Axial location	T1/ T2WI	Size (mm)	Dilated vessels	Cord edema	Syrinx	CE	Others
1	M	46	Myxopapillary ependymoma	LBP/RLP	Central	Low /Hetero	28	No	extensive	Yes	Yes	Fluid-fluid level in cyst
2	M	61	Hemangioblastoma	LBP	Mainly central	Iso /low	25	No	extensive	Yes	Yes	Eccentric daughter nodule
3	M	31	Ventriculus Terminalis	LBP/RLP Urinary disturbance	Central	Low /High	21	No	none	No	Yes	Internal septa Peripheral rim CE
4	F	35	Dermoid cyst	LBP/Urinary disturbance	Central	High /Hetero	62	No	none	No	Yes	Irregular wall thickening
5	F	37	Hemangioblastoma	LBP/buttock pain	Eccentric	Low /High	50	Yes	extensive	No	Yes	Eccentric daughter nodule
6	M	59	Spinal AVF	LE weakness /sensory change	Central	Low /High	48	Yes	extensive	Yes	Yes	Patchy enhancement of the conus medullaris

Note.— M=male; F=female; LBP=lower back pain; RLP=right leg pain; AVF=arteriovenous fistula; CE=contrast enhancement

RESULTS

Six intramedullary spinal masses were located from the 12th level of the thoracic vertebra to the 2nd level of the lumbar vertebra and their size varied from 21 to 62 mm (Table 1). Three cystic masses were intermingled with the solid portion, which showed variable signal intensities on T1 weighted (T1WI) and T2-weighted images (T2WI) (Figs. 1, 3). Two purely cystic lesions showed the same signal as the cerebrospinal fluid (CSF) (Fig. 2). Contrast enhancement was seen in all intramedul-lary masses. An eccentric enhancing nodule was noted in two hemangioblastomas (Figs. 1a, 2b) and unusual peripheral rim enhancement with septation was seen in ventriculus terminalis (Fig. 2a). Patchy enhancement of the CM was observed in spinal arteriovenous fistula (AVF) (Fig. 1b). Extensive cord edema and hydrosyrinx was seen adjacent to the intramedullary lesions in three cases. Vascular signal voids were found in two hemangioblastomas and one spinal AVF which were confirmed as "dilated medullary vein" on spinal angiography. On the follow-up two cases (patient 1 and 2), which showed residual or recurrent contrast- MR imaging, four cases showed almost complete resolution after total surgical resection or embolization and enhancing lesions two years after surgery.

Spinal angiography was performed in three patients to evaluate preoperative embolization of two hemangioblastomas and one AVF and transarterial embolization was performed in the latter.

Five patients had frequent lower back pain, combined with leg pain in two and buttock pain in one. Urinary disturbance was seen in two patients. In spinal AVF, sudden-onset weakness in extremities and sensory change was noted. All patients improved their symptoms after operation or embolization.

DISCUSSION

We described six different cases of intramedullary

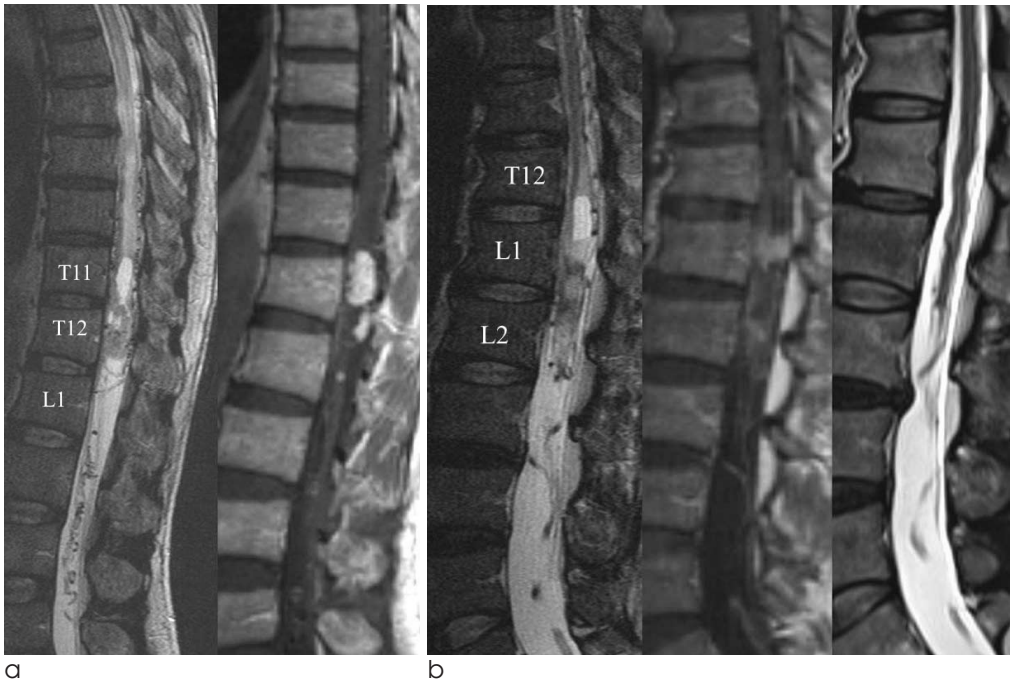

Fig. 1. Intramedullary hemangioblastoma (**a**) versus Spinal arteriovenous shunt (**b**).
a. T2-weighted sagittal image (left) shows solid mass of the conus medullaris with hydrosyrinx, extensive cord edema and dilated medullary veins. Contrast enhanced T1-weighted sagittal image (right) shows enhancement of two solid components. **b.** T2-weighted sagittal image (left) shows swelling and high signal intensity and atrophy of the conus medullaris with hydrosyrinx, dilated medullary vein and proximal cord edema. Contrast enhanced T1-weighted sagittal image (middle) shows focal enhancement of the conus medullaris, probably due to chronic ischemic insult from venous hypertension. On follow-up MR (right) cord edema and syrinx was disappeared after embolization.

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spinal pathologies involving the CM, including ependymoma, hemangioblastomas, dermoid cyst, ventriculus terminalis and spinal AVF. Irrespective of tissue pathology, intramedullary lesions involving the CM had some MR imaging in common; 1) relatively larger mass, more than 20 mm with swelling of the

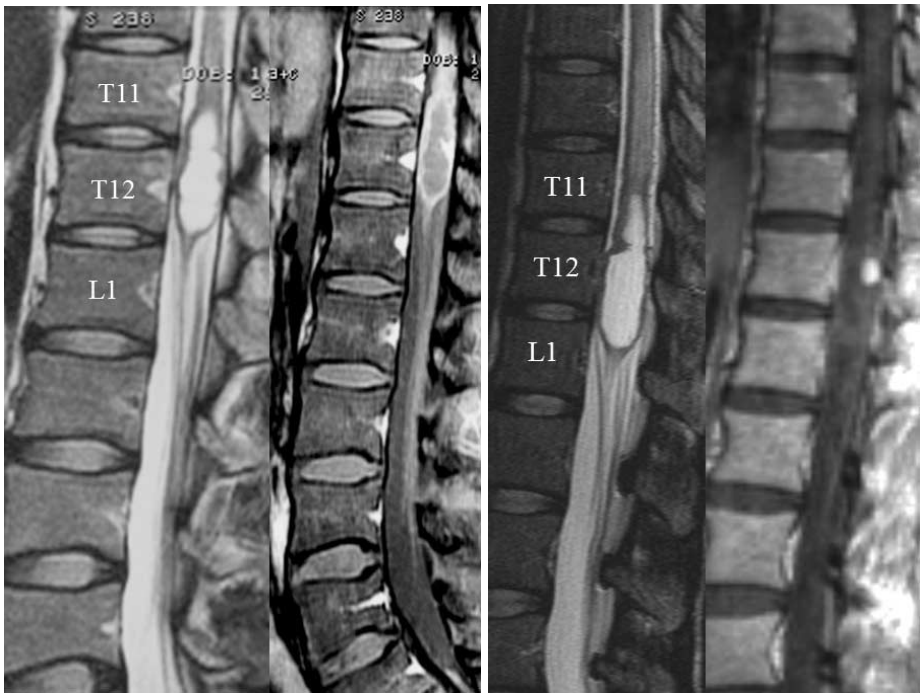

Fig. 2. Ventriculus terminalis (**a**) versus Intramedullary hemangioblastoma (**b**).
a. T2-weighted sagittal image (left) shows cystic mass in the conus medullaris without hydrosyrinx and cord edema. Contrast enhanced T1-weighted sagittal image (right) shows peripheral rim enhancement of the cyst with internal septum. **b.** T2-weighted sagittal image (left) shows cystic mass of the conus medullaris with extensive cord edema and dilated perimedullary vein. Contrast enhanced T1-weighted sagittal image (right) shows eccentric enhancement of the nodular component within the intratumoral cyst.

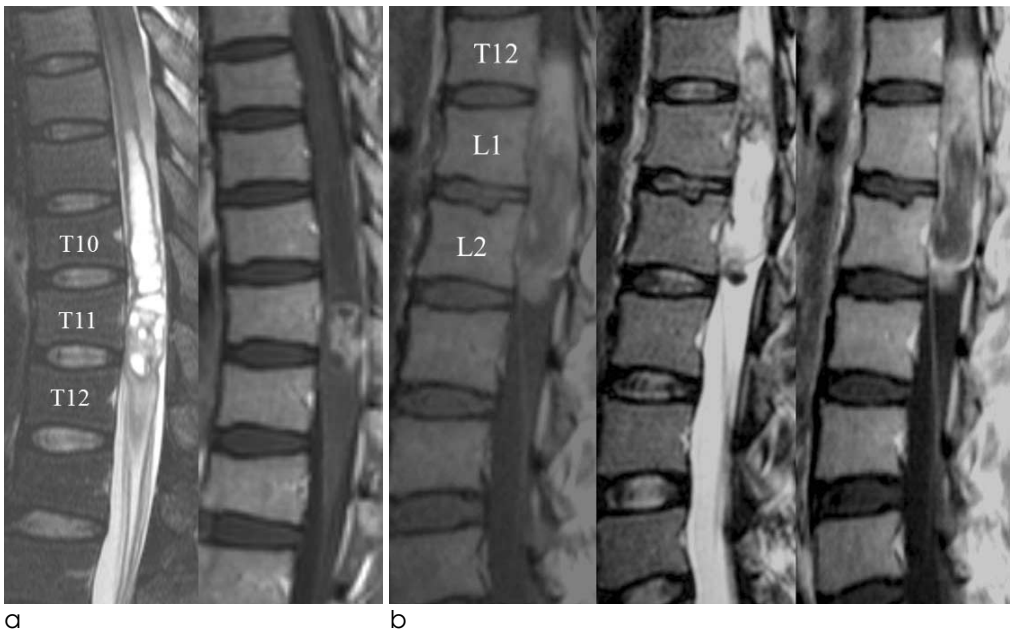

Fig. 3. Myxopapillary ependymoma (**a**) versus Dermoid cyst (**b**).
a. T2-weighted sagittal image (left) shows solid and cystic mass in the conus medullaris with extensive hydrosyrinx and cord edema. Contrast enhanced T1-weighted sagittal image (right) shows enhancement of the solid component. **b.** T1-weighted sagittal image (left) shows heterogeneous signal intensity within the intratumoral cyst and T2-weighted sagittal image (middle) shows solid and cystic mass of the conus medullaris without hydrosyrinx. Contrast enhanced T1-weighted sagittal image (right) shows rim enhancement of the solid component.

CM, 2) contrast enhancement of the solid component, 3) central location on the CM except one hemangioblastoma. The unusual MR features in this study were syringomyelia and patch cord enhancement in spinal AVF, cystic wall enhancement with an internal septum in the ventriculus terminalis and peripheral rim enhancement of the dermoid cyst.

Tumor size of our five cases involving the CM was relatively large, more than 20 mm. Epstein et al. (17) suggested dominant motor symptoms were commonly associated with very large ependymomas and a poorer postoperative outcome secondary to the increased surgical risk. Although swelling of the CM was found, hydrosyrinx occurred in only three cases, two cases of which were combined with extensive cord edema and dilated medullary vein. The cause of syrinx and cord edema may be associated with intramedul-lary cord tumor as well as venous hypertension. While the syrinx was frequently reported in 9–50% of ependymoma (16, 18) and 40–81% of hemangioblastoma (19–22), the syrinx was a rare pathology in case of spinal AVF. Srivatanakul et al. (11) reported that venous hypertension in the spinal cord was the trigger for the development of syringomyelia and showed disappearance of syrinx after transarterial embolization.

Typical MR imaging of spinal ependymomas usually show isointensity or slightly hypointensity on unenhanced T1-weighted images (15, 18, 23, 24). Ependymomas show hyperintencity in rare occasions, when the myxopapillary ependymoma is associated with hemorrhage or mucin production (18, 23). After gadolinium enhancement, lesions show homogenous, heterogenous or only rim enhancement (15, 18, 23, 24). About 20% of cases of ependymomas can show low signal intensity rim on T2-weighted images, which indicates the presence of hemosiderin deposit (23). Combined nontumoral cysts are common in about 78-84% ependymomas and incidence of tumoral cysts are about 4–50%, which is variable (16, 18, 23). Syringomyelia were variably associated in about 34–90% of ependymomas (18, 25). Our one case of ependymomas also showed cystic portion, which is also probably related to hemorrhage or necrosis.

Spinal hemangioblastomas shows hypointense to isointense on T1WI and isointense to hyperintense on T2WI (20, 26–28). On T2WI, hemangioblastomas can have intermixed focal flow voids (20, 26–28). Cyst formation or syringohydromyelia is very common (19, 20, 22, 26, 29) like our two cases. Our two cases of hemangioblastomas showed mostly cystic lesions with eccentric enhancing nodules which was compatible with typical MR findings of the previous studies (26, 28, 30). Both cases showed extensive cord edema and flow-void signal within the lesion.

The ventriculus terminalis is a round cystic cavity with smooth wall, no internal septa and no contrast enhancement of the cyst or its wall (31–38). However, there was an internal septation and cystic wall enhancement in our case, which would lead to surgical resection because of misdiagnosis as spinal astrocytoma. Cystic wall enhancement was likely to be originated from venous hypertension or combined inflammatory change.

Spinal dermoid cysts showed usually hyperintensity on T1WI due to fatty secretion of sebaceous gland and cholesterol and usually hypointensity on T2WI but might be homogenous or heterogenous (39, 40). Spinal dermoid cysts did not usually enhance on post-contrast examination (3, 41). However our case showed heterogeneous intensity in T1 and T2WI as well as peripheral enhancement of the solid component (Fig. 3b). Enhancement of spinal dermoid cysts was a rare entity; only two cases were reported that was peripherally enhanced (42, 43).

Typical MR imaging of dural AVF show increased signal intensity on T2WI throughout the central area of the cord, particularly at CM and decreased signal intensity on T1WI. Prominent signal voids are frequently associated in subarachnoid space, which are the result of dilated vein. Spinal angiography provides the exact location and size of a lesion and information about feeding and draining vessels. Our case showed mostly cystic lesion with expansive cord edema and focal enhancement, probably due to chronic ischemic insult.

In conclusion, although this study had some limitation of heterogeneity and small numbers, intramedullary spinal pathologies of the CM have typical MR characteristics as well as some exceptional imaging features. In evaluation of intramedullary spinal lesions in the CM, it is necessary to consider these unusual MR findings and discriminate various pathologies with prudence and caution.

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척수 원추부에 발생한 척수내 병변: 자기공명영상을 이용한 감별 진단

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목적: 척수 원추부에 국한된 척수내 병변은 현재까지 보고된 바가 드물다. 이 논문에서는 척수 원추부에 국한된 척추내 병변인 상의세포종, 혈관모세포종, 유피낭종, 종말 뇌실, 척추 동정맥류 등 6 증례의 자기공명영상의 다양한 소견을 보고하고 감별점을 논의해 보고자 한다.

대상과 방법: 2004년부터 2010년까지 본원의 자료를 바탕으로 하여 총 6명의 환자 (남:여 = 4:2, 평균연령 = 44.3세)가 대상이 되었으며 이들의 방사선학적소견과 임상증상 등을 각각 검토하였다. 모든 환자들은 수술 전 후 가돌리늄 조영증강 자기공명영상을 시행하였다. 그리고 종양 크기, 위치, 자기공명 신호강도, 혈관성 유동공백의 유무, 관 혹은 낭의 존재 유무, 부종의 존재 유무, 조영증강 양상에 따라 영상을 비교하였다.

결과: 조영 증강은 모든 척수내 병변에 나타났다. 2 증례의 혈관모세포종에서 편심적 조영증강 결절이 보였으며 종말 뇌실 증례에서 흔치 않은 테두리 조영증강과 격막이 보였다. 척추 동정맥류에서는 반점형 조영증강이 관찰되었다. 3개의 증례에서 척수내 병변에 근접하여 광범위한 척수 부종과 수관이 보였다. 2 증례의 혈관모세포종과 1 증례의 척추 동정맥류에서 혈관성 유동공백이 관찰되었다.

결론: 척수 원추부에 있는 척수내 질환을 평가하는데 있어 이러한 드문 자기공명영상의 특징과 다양한 병리를 신중하게 고려해야 할 필요가 있다.

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