Case Report

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Intradural Extramedullary Ependymoma with Hemorrhage: A Case Report

출혈을 동반한 경막내 수외 뇌실막세포종: 증례 보고

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In adults, spinal ependymomas are usually found in intramedullary locations. However, intradural extramedullary spinal ependymomas are rare. Additionally, spinal ependymomas usually show iso to hypointensity on T1-weighted images without hemorrhage. Herein, we present a rare case of a 43-year-old female with a pathologically confirmed intradural extramedullary ependymoma that showed hyperintensity on T1-weighted imaging accompanied by hemorrhage.

Index terms Spinal Cord Neoplasms, Intradural-Extramedullary; Ependymoma; Magnetic Resonance Imaging

INTRODUCTION

Spinal ependymomas are usually located in the intramedullary compartment in adults and account for 60% of all intramedullary lesions. Spinal ependymomas are generally well-circumscribed primary central nervous system tumors. MR findings mostly show iso-to-hyperintensity on T2-weighted images and hypo-to-isointensity on T1-weighted images. These tumors demonstrate marked but often heterogeneous enhancement and cyst formation is common (1, 2). Although these tumors are often benign, a few may follow a malignant course (3).

Ependymomas that arise outside the conus medullaris, cauda equina, and terminal filum or originate from ectopic ependymal cells are unusual. Intradural extramedullary (IDEM) ependymomas are extremely rare. The most common primary IDEM neoplasms are menin-

giomas and schwannomas followed by neurofibromas. Less common neoplasms include solitary fibrous tumors, hemangiopericytomas, and malignant peripheral nerve sheath tumors.

To our knowledge, no reported article has described an IDEM ependymoma with a high signal intensity on T1-weighted images. In this report, we present the clinical, radiological, and pathological features of an IDEM ependymoma showing high intensity on T1-weighted imaging, with perilesional hemorrhage.

CASE REPORT

A 43-year-old female visited our institution with radiating pain in both buttocks, posterior thighs, and calves that had started 1 year prior to presentation. She denied a history of trauma.

Lateral thoracolumbar spine radiography confirmed intervertebral disc space narrowing at the L5–S1 level was confirmed. L-spine CT revealed an intradural, mildly hyperdense mass at the T12–L1 level without calcification (Fig. 1A).

Spinal MRI revealed an intradural lesion that displaced the conus medullaris anteriorly and to the right. The IDEM mass was approximately 2.5 cm in length and located at the T12–L1 level near the conus medullaris. The mass showed high signal intensity on both T1- and T2-weighted imaging. After gadolinium injection, the lesion showed mild contrast enhancement (Fig. 1B, C). The preoperative diagnosis was a schwannoma with hemorrhage. The possibility of meningioma, myxopapillary ependymoma, or metastasis was not excluded. Contrast-enhanced brain MR images were not obtained before and after surgery, so it was difficult to differentiate whether the finding was due to metastasis. However, metastasis is unlikely, because it is often accompanied by edema of the surrounding spinal cord.

T12–L1 left hemilaminectomy and tumor removal were performed under general anesthesia. A mass located in the intradural space was found intraoperatively. Hemorrhage was prominent in the operative field. No postoperative complications were noted. Six days after tumor removal, follow-up MRI revealed no demonstrable tumor recurrence or remnant lesions (Fig. 1D).

Histological analysis revealed diffuse hemorrhagic necrosis with hemosiderin-laden macrophages and granulation tissue with a viable focal tumor (Fig. 1E). The tumor was composed of isomorphic round to oval nuclei arranged apart from vessels, forming a pseudorosette. A few nuclei were clear, but most showed speckled chromatin. No tumor necrosis, mitosis, or microvascular proliferation was observed. Myxoid changes or microcysts were not observed (as observed in myxopapillary ependymomas). Immunohistochemical staining revealed glial fibrillary acidic protein and vimentin positivity, epithelial membrane antigen dot-like staining, cytokeratin, and SRY-related HMG-box 10 (SOX 10) negativity. The Ki-67 proliferation index was less than 1%.

This study was approved by the Institutional Review Board of our institution, which waived the requirement for informed consent (IRB No. 2022-12-057).

DISCUSSION

IDEM ependymomas are much rarer than intramedullary ependymomas are. The mecha-

nisms underlying the development of IDEM ependymomas are not well understood. In certain studies, these lesion-type tumors were thought to arise from ectopic ependymal cell rest. Some ependymal cells may remain inside during neural tube closure (3).

Fig. 1. A 43-year-old female with spinal intradural extramedullary ependymoma with hemorrhage.
A. Radiograph shows intervertebral disc space narrowing at the L5–S1 level (left). CT scans with a soft tissue window shows an intradural mildly hyperdense mass (arrows) at the T12–L1 level. The mass had no internal calcifications.
B. Sagittal T1-weighted (left), T2-weighted (middle), and postcontrast T1-weighted MRI (right) show the intradural extramedullary tumor at T12–L1. The mass shows high signal intensity on T1- and T2-weighted images in the dorsal intradural extramedullary space, from T12 to L1. The tumor exhibits mild contrast enhancement.
C. Axial T1-weighted (left), T2-weighted (middle), and post-contrast T1-weighted MRI (right). The mass had displaced the spinal cord on the right side. The mass shows high signal intensity on T1- and T2-weighted images on the dorsal side of the intradural extramedullary space from T12 to L1. The tumor exhibits mild contrast enhancement.

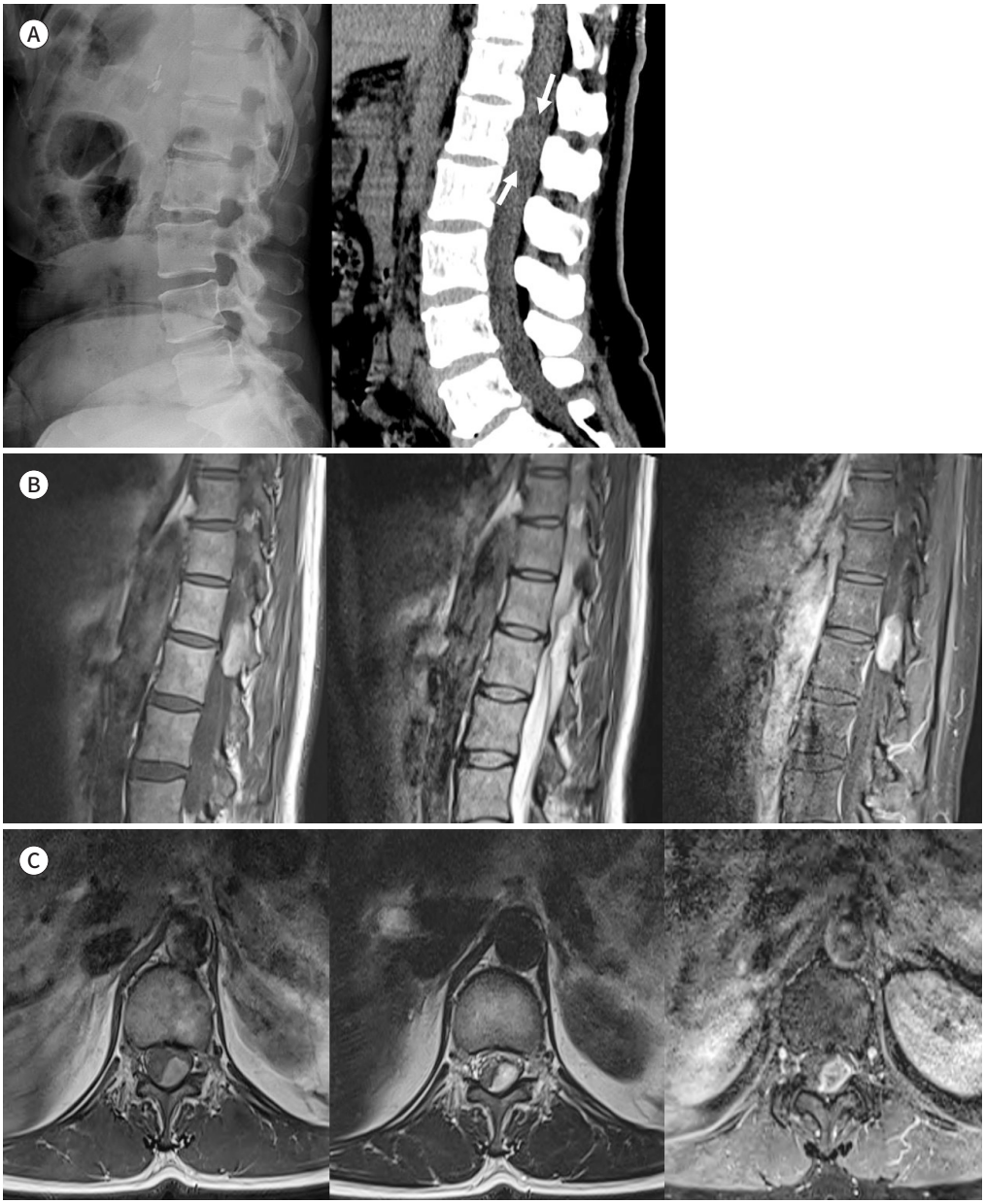
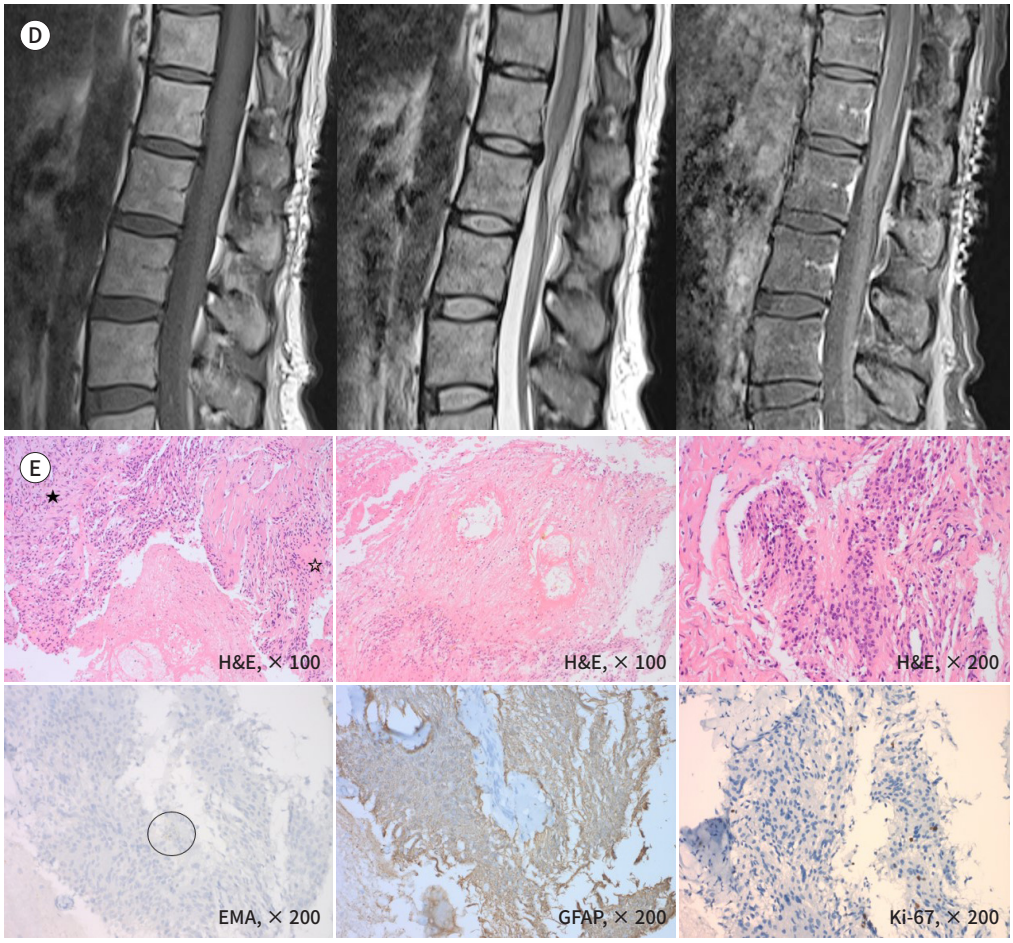


Fig. 1. A 43-year-old female with spinal intradural extramedullary ependymoma with hemorrhage.
D. After tumor removal, postoperative sagittal T1-weighted (left), T1-weighted (middle), and post-contrast T1-weighted MRI (right). After tumor removal, follow-up MRI reveals no demonstrable tumor recurrence or remnant lesions.
E. Microscopic findings of the spinal ependymomas. A scattered viable tumor (white star) reveals an anucleated zone and a cellular region. Focal inflamed granulation tissue (black star) and diffuse necrosis (center) are observed in the background (upper left). Hemorrhagic necrosis shows central vascular necrosis and hemosiderin-laden macrophages with a viable fibrillary glial matrix in the periphery (upper middle). Ependymoma shows pseudorosettes composed of isomorphic round nuclei, apart from the blood vessels (upper right). The EMA staining shows is focal cytoplasmic dot-like staining (circle, lower left). GFAP-stained perivascular radiating tumor process (lower middle) are also seen. A very low proliferation index was identified using Ki-67 staining (lower right).
EMA = epithelial membrane antigen, GFAP = glial fibrillary acidic protein, H&E = hematoxylin and eosin stain



Similar to typical intradural ependymomas, IDEM ependymomas occur irrespective of age, but are most common in the third to fifth decades of life. The overall prevalence of the majority of intradural ependymomas among males and females is equal, but in IDEM ependymomas, there appears to be a female preponderance. Most cases of IDEM ependymomas present with progressive myelopathy, as well as pain and weakness, according to the tumor location (3). A similar presentation was observed in the present case. Some features commonly associated with IDEM ependymomas include diagnosis in the fifth decade, female predominance, thoracic spinal location, encapsulated lesions not attached to the central ner-

vous system, and lack of other tumors noted in the brain or spinal cord (4).

Differential diagnosis of IDEM masses with T1 high-intensity signals includes schwannoma with hemorrhage, meningioma, paraganglioma, myxopapillary ependymoma, and cancer metastasis. Cancer metastasis can be differentiated owing to its multiplicity. Our initial differential diagnosis included schwannoma with hemorrhage.

Schwannomas are benign nerve sheath tumors that are more common outside the CNS and usually involve peripheral nerves and subcutaneous tissue. Interestingly, solitary and sporadic lesions were the most common (95%). MRI of schwannomas classically reveals well-circumscribed T1 iso-/hypointense and T2 hyperintense nodular intradural mass (5). Schwannomas are frequently associated with hemorrhage, intrinsic vascular changes, cystic changes, and fatty degeneration.

Paragangliomas are usually isointense to the conus medullaris on T1WI but might be hypointense. On T2WI, paragangliomas are hyperintense and sometimes heterogeneous, owing to subacute blood clots. Serpentine flow voids are a frequent feature attributed to the hypervascularity of the lesion or to congested veins, which are compressed by the mass (6).

Myxopapillary ependymomas are present when they become larger and sausage-shaped, spanning more than one vertebral level, but small oval tumors are also observed. They tend to be hypointense to isointense compared to the intensity of the normal spinal cord on T1WI, and hyperintense on PD-weighted imaging and T2WI. Signal intensity may also be affected by blood products, and calcification may lead to regions of low T2 signal intensity (6).

Previously reported IDEM ependymomas are usually iso-to-hypointense on T1-weighted images and iso-to-hyperintense on T2-weighted images with homogeneous enhancement after gadolinium injection (1, 7-9). To date, fewer than 30 cases of IDEM ependymoma have been reported. Most showed T1 isointensities (Supplementary Table 1 in the online-only Data Supplement).

Kim et al. (9) reported a case of IDEM epedymoma that showed mild hyperintensity on T1-weighted imaging. In their case report, they regarded partial hemorrhage as the cause of hyperintensity on T1-weight imaging. In our case, diffuse hemorrhagic necrosis with hemosiderin-laden macrophages was observed on pathologic review. Hemorrhagic necrosis was considered the cause of hyperintensity on T1-weighted imaging.

Shin et al. (10) reported typical and atypical MR findings of intramedullary spinal ependymomas, including T1-weighted and T2-weighted images, and contrast-enhanced morphology in 47 cases of intramedullary ependymoma. Out of a total of 47 cases, only three (6%) showed high signal intensity on T1 images. Even in intramedullary ependymomas, a high signal intensity on T1-weighted imaging is relatively rare. Most cases showed T1-iso signal intensity (81%) and T2-high signal intensity (72%).

The definitive treatment of choice for IDEM ependymomas is gross total resection. Small tumors can be removed en bloc; however, larger tumors may require intracapsular decompression. Additionally, hemilaminectomy and tumor removal were performed in our patient. Most spinal IDEM ependymomas are described as benign lesions; however, recurrence, anaplastic transformation, and metastasis can occur. Therefore, close follow-up should be performed after complete resection because of the possibility of recurrence, metastasis, and anaplastic transformation (3).

In conclusion, although ependymomas are uncommon, the possibility of ependymoma should be considered when an IDEM spinal tumor develops. Additionally, it is important to know that IDEM ependymomas may show hyperintensity on T1-weighted images.

Supplementary Materials

The online-only Data Supplement is available with this article at http://doi.org/10.3348/jksr.2022.0173.

Author Contributions

Conceptualization, L.S.J.; data curation, P.S.H., C.H.J.; formal analysis, L.S.J.; supervision, L.S.J.; visualization; C.C.; writing—original draft, C.C.; and writing—review & editing, L.S.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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출혈을 동반한 경막내 수외 뇌실막세포종: 증례 보고

최창원[1] · 이선주[1]* · 팽성화[2] · 조화진[3]

뇌실막세포종은 일반적으로 성인의 척수내 위치에서 발견된다. 경막내 수외 뇌실막세포종은 드물다. 또한, 뇌실막세포종은 일반적으로 출혈 없이 T1 강조영상에서 등- 또는 저신호강도를 나타낸다. 저자들은 43세 여성에서 발생한 드문 출혈을 동반하며 T1 강조영상에서 고신호강도를 보인 경막내 수외 뇌실막세포종으로 확진된 1예를 보고하고자 한다.

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