



Non-Enhancing Intradural Extramedullary Ependymoma: A Case Report

조영증강이 되지 않는 경막내 수외 뇌실막세포종:
증례 보고

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Spinal ependymomas are generally located in the intramedullary compartment in adults. Intradural extramedullary spinal ependymomas are extremely rare. Spinal ependymomas show various contrast enhancements on MRI. In this study, we report a rare case of a 52-year-old female who had a pathologically confirmed intradural extramedullary ependymoma that showed no enhancement on MRI.

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

INTRODUCTION

Spinal ependymoma is generally found in the intramedullary location in adults, accounting for 60% of the total intramedullary lesions (1). Intradural extramedullary ependymoma is relatively rare (2-4). Most spinal ependymomas have well-defined and intense contrast enhancement after the injection of contrast medium (5-7). This case report focuses on the unusual MR finding of intradural extramedullary ependymoma at conus medullaris without contrast enhancement.

CASE REPORT

A 52-year-old female presented at the hospital in March 2020 with low back pain with direct tenderness and both leg weakness. Neurologic examination revealed bilateral paraparesis grade IV. The results of Deep tendon reflexes and Patrick straight leg raise tests were normal.

Degenerative spondylosis was confirmed through lateral thoraco-lumbar (TL) spine




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radiograph, and wedge-shaped T11 vertebral body was incidentally found (Fig. 1A). TL spine CT revealed an intradural iso-dense mass at the T12–L1 level without calcification. The incidentally founded wedge-shaped T11 vertebral body was a butterfly vertebra, normal variation (Fig. 1A). The results of spinal MRI demonstrated an intradural lesion, posteriorly located and compressing the conus medullaris anteriorly. There was an enlargement of cerebrospinal fluid (CSF) space adjacent to the mass. The intradural extramedullary mass was approximately 3.9 cm in length and was located at the T12–L1 level near conus medullaris.

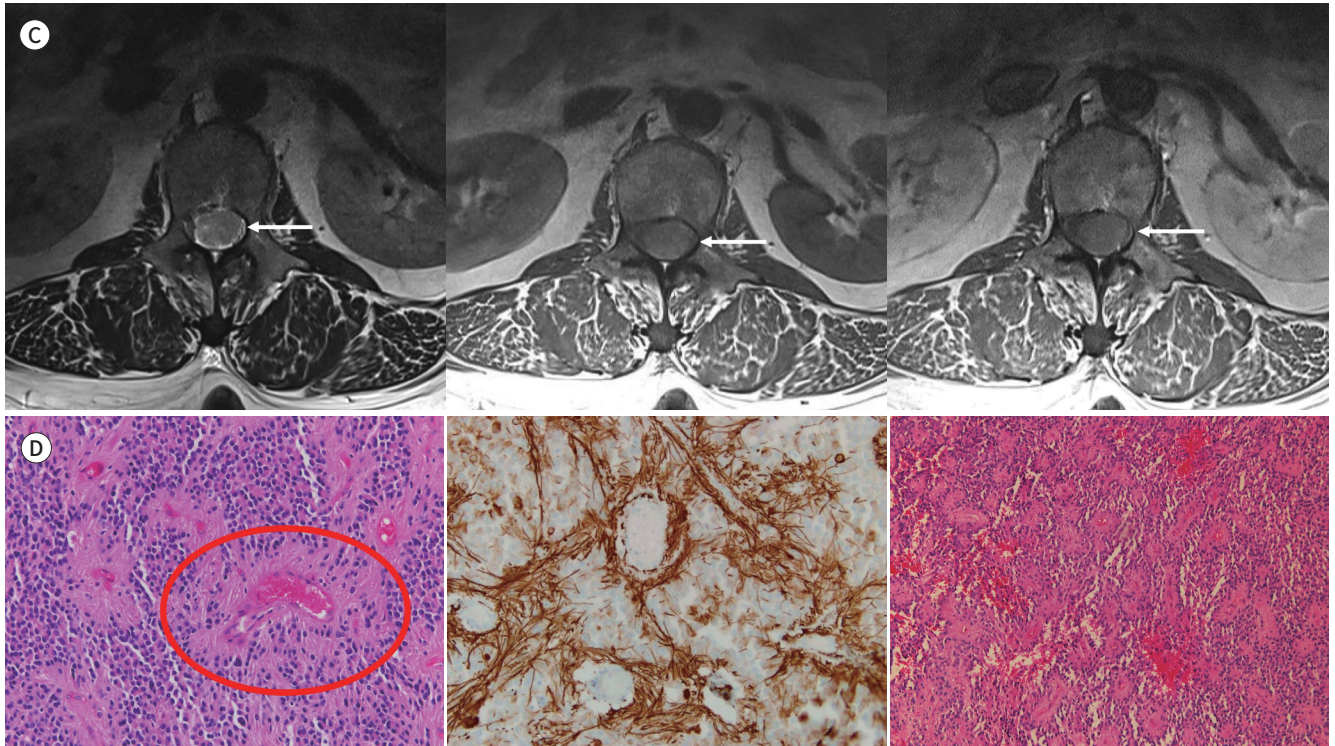
Fig. 1. A 52-year-old female with spinal intradural extramedullary ependymoma.

A. Radiograph shows degenerative spondylosis and incidentally found butterfly vertebra at T11 (left). Sagittal CT scans with a soft tissue window (middle) and bone window (right) show the intradural isodense mass (arrows) at the T12–L1 level. The mass has no internal calcification.
B. Sagittal T2-weighted (left), T1-weighted (middle), and postcontrast T1-weighted MRI (right) show the intradural extramedullary tumor at T12–L1 (white arrows). The central portion of the mass shows high signal intensity on the T2-weighted image (black arrow).



Fig. 1. A 52-year-old female with spinal intradural extramedullary ependymoma.

C. Axial T2-weighted (left), T1-weighted (middle), and postcontrast T1-weighted MRI (right). The mass (arrows) shows no contrast enhancement.
D. Histologic feature of ependymoma. Ependymal pseudorosettes (red circle) are composed of tumor cells arranged around blood vessels (hematoxylin and eosin stain, $\times 200$) (left). Pseudorosettes are positive for glial fibrillary acidic protein (GFAP, $\times 200$) (middle). Ependymoma shows partial internal hemorrhage (hematoxylin and eosin stain, $\times 100$) (right).



The mass showed a slightly high signal intensity on both T1 and T2-weighted images. The central region of the mass had a higher signal intensity on T2-weighted image than in the peripheral region. After gadolinium injection, the lesion showed no contrast enhancement (Fig. 1B, C). The preoperative diagnosis was schwannoma. The possibility of hematoma and myxopapillary ependymoma was not excluded.

T12-L1 total laminectomy and dural incision were performed under general anesthesia. The white round well-defined mass located in the intradural space was found intraoperatively. A strand of the nerve was attached to the mass, and it was well separated. Post-operative complications were not developed.

The results of histologic examination showed perivascular pseudorosettes. Immunohistochemical staining was performed and positive for glial fibrillary acid protein (GFAP). Additionally, partial hemorrhage was observed (Fig. 1D). The final histologic diagnosis was a World Health Organization grade II ependymoma.

DISCUSSION

Intradural extramedullary ependymoma is much rarer than intramedullary ependymoma. The mechanism underlying the occurrence of intradural extramedullary ependymoma remains unknown (1). Overall, two hypotheses for the ectopic ependymoma have been pro-

posed (8, 9). To date, only nine cases of intradural extramedullary ependymoma have been reported, and all of them had intense contrast enhancement. All patients, including the one reported in the present study, were female. Accordingly, the hypothesis has been formulated that hormonal mechanism may have contributed to the development of the tumor, which seems reasonable in our case (9). Additionally, previous studies have also discussed the possibility that the location of these exceptional tumors is caused by a heterotopic glial cell in the intradural extramedullary space (8, 9).

Previously reported intradural extramedullary ependymomas were usually iso-to-hypointense on T1-weighted image and hyperintense on T2-weighted image, with a homogeneous enhancement after gadolinium injection (3, 4). However, none of the reported cases showed image findings similar to those observed in our patient. Intramedullary ependymoma can be accompanied by cystic changes and may not be enhanced after the contrast medium injected (5-7). However, in our case, it was an intradural extramedullary solid tumor without cystic change, and there was no contrast enhancement. Non-enhancing ependymoma without cystic change was reported among pediatric brain tumors (5). Only three cases of intramedullary ependymoma without contrast enhancement were reported in the literature (10). In addition, on contrast-enhanced T1-weighted image, the signal intensity of this tumor was slightly higher than that of CSF and muscle. We considered the reason for hyperintensity on T1-weighted image as partial hemorrhage.

In conclusion, ependymoma should be considered when we encounter intradural extramedullary spinal neoplasm, even if it is not common. Furthermore, it is important to know that intradural extramedullary ependymoma may not be enhanced after the injection of contrast medium.

Author Contributions

Conceptualization, K.H.; data curation, K.H., K.J.; formal analysis, K.H., K.J.; funding acquisition, K.H.; investigation, K.H., K.J.; methodology, K.H., K.J.; project administration, K.H.; resources, K.H., K.J.; software, K.H., K.J.; supervision, K.H.; validation, K.H.; visualization, all authors; writing—original draft, K.H., K.J.; and writing—review & editing, K.H., K.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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조영증강이 되지 않는 경막내 수외 뇌실막세포종: 증례 보고

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척수 뇌실막세포종은 일반적으로 성인에서 척수 내에 위치한다. 경막내 수외 척수 뇌실막세포종은 극히 드물다. 대부분의 척수 뇌실막세포종은 MRI에서 다양한 조영증강을 보인다. 저자는 MRI에서 조영증강을 보이지 않으며, 병리학적으로 확인된 52세 여자 환자의 경막내 수외 뇌실막세포종을 경험하여 이에 대해 보고하고자 한다.

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