Primary Extramedullary Ependymoma of the Cervical Spine: Case Report and Review of the Literature

Dong Wuk Son, M.D., Geun Sung Song, M.D., In Ho Han, M.D., Byung Kwan Choi, M.D.
Department of Neurosurgery, School of Medicine, Pusan National University, Busan, Korea

Intradural extramedullary (IDEM) ependymomas occur very rarely and little has been reported about their clinical characteristics. The authors present a case of a 57-year-old woman with an IDEM ependymoma. She was referred for the evaluation of a 4-month history of increasing neck pain and muscular weakness of the left extremities. Magnetic resonance imaging (MRI) of the cervical spine demonstrated an IDEM tumor with spinal cord compression. At the time of surgery, an encapsulated IDEM tumor without a dural attachment or medullary infiltration was noted, but the tumor capsule adherent to the spinal cord and root was left in place to minimize the risk of neurological sequelae. Histologic examination revealed a benign classic ependymoma. The post-operative course was uneventful and radiotherapy was performed. The patient showed an excellent clinical recovery, with no recurrence after 5 years of follow-up.

Key Words: Intradural extramedullary, Ependymoma, Cervical spine.

INTRODUCTION

Ependymomas are the most common tumors of the spinal cord in adults, comprising up to 60% of the tumors found at this location. Because ependymomas arise from ependymal cells located in the central canal of the spinal cord, ependymomas are completely positioned within the cord and are rarely found outside it, excluding myxopapillary ependymomas. Intradural extramedullary (IDEM) ependymomas are very rare and usually not considered in the differential diagnosis of IDEM spinal tumors. Recently we experienced a case of IDEM ependymoma of the cervical spine. In this report, we discuss the clinical findings, radiologic features, surgical management and prognosis of this rare tumor.

CASE REPORT

A 57-year-old woman was admitted with increasing neck pain and muscular weakness of the left extremities for 4 months. The neurologic signs consisted of left hemiparesis (upper grade IV/ lower grade IV), hypesthesia below dermatome C3 and paresthesias of the left extremities. Biceps and triceps reflexes of the left side were +2/+3, knee jerk and ankle jerk were +2/+3. The voiding sense and anal sphincter tone were intact.

Magnetic resonance imaging of the spine showed a large intradural extramedullary mass extending from C2-C6 with spinal cord compression. The lesion exhibited iso to high signal intensity on T2-weighted images and iso signal intensity on T1-weighted images with mild enhancement after gadolinium injection (Fig. 1).

The pre-operative diagnostic impression was a neurinoma, neurofibroma or meningioma. At the time of surgery, a posterior laminotomy from C2-C6 was performed. When the dura mater was opened, a dark-pinkish, extramedullary encapsulated tumor was observed (Fig. 2). The compressed spinal cord was displaced to the right. The tumor was not in continuity with the spinal cord, dura, or rootlets, although the lesion was adherent to the spinal cord and rootlets. Under an operating microscope, we dissected the arachnoid around the tumor and removed the tumor without damage to the medulla. An attempt was made to mobilize the tumor capsule from the spinal cord and nerve roots, but it seemed to be so dangerous because of the firm adhesion to some part of spinal root and cord that we left the adherent portion of tumor capsule in place to minimize the risk of neurological sequelae. Bilateral laminoplasty from C2-C6 was performed at the end of the surgery.

Histologic examination revealed a densely cellular glial tumor. The tumor cells were characterized by round, oval nuclei, moderate hyperchromasia and eosinophilic cytoplasm. Perivascular pseudorosettes were noted (Fig. 3). The cells were immu-
noreactive for glial fibrillary acidic protein (GFAP), but negative for epithelial membrane antigen (EMA) and reticulin. There were no mitosis or anaplastic changes. Ki-67 indicated a mitotic index under 4%. These findings were consistent with a benign ependymoma, WHO Grade II.

Post-operatively, the patient's neurologic condition improved. No other lesions were detected on the brain and spinal MRI. Post-operative radiotherapy therapy was performed [1.8 Gy in 28 fractions (total, 50.4 Gy)]. Six months after surgery, the neurologic function had recovered to near-normal. There were no signs of local recurrence and distant dissemination on magnetic resonance imaging after 5 years of follow-up (Fig. 4).

DISCUSSION

Although ependymomas are the most common gliomas of the spinal cord, conus medullaris or filum terminale in adults, the intradural extramedullary location of this glial lesion is exceptional. Few cases of purely IDEM ependymomas have been reported in the literature\textsuperscript{1-11,13,15}.

The precise histogenesis of IDEM ependymomas is unclear yet. Ependymomas usually arise from the ependymal cells that form the lining of the ventricles and central canal of the spinal cord. However, IDEM ependymomas may arise from heterotopic ependymal cell rests pinched off from the neural tube during its closure\textsuperscript{2,4,7,9,15}. This hypothesis is supported by the following Cooper's descriptions\textsuperscript{4}: lack of an apparent attachment to the central nervous system and absence of signs of a primary neoplastic process within the brain or spinal cord, the encapsulated appearance, location along the neuraxis and frequent association with congenital anomalies.

As with typical intradural ependymomas, IDEM ependymomas occur irrespective of age, but are most common in the third to fifth decades of life\textsuperscript{1-11,13,15}. The overall prevalence of the majority of intradural ependymomas among males and females is equal\textsuperscript{12}, but, in IDEM ependymomas, there appears to be a female preponderence\textsuperscript{3-11,13,15}. Therefore, a hormonal mechanism is thought to play a role in the development of this rare tumor\textsuperscript{5,10} but further studies are required to clarify its pathogenesis\textsuperscript{7}. Most spinal IDEM ependymomas affect the thoracic spine\textsuperscript{5,10} in contrast to intramedullary ependymomas, mainly located at the cervical spinal cord and the conus medullaris re-
Generally, the presentation of the IDEM ependymomas is not different from that of more common intradural extramedullary lesions\(^4\). A history of pain, and a progressive history of medullary compression are usually reported. Our case also presented with pain and corticospinal tract dysfunction.

Imaging findings are non-specific and show a tumor located within the intradural extramedullary space. In the literature, they were usually homogenously enhanced after gadolinium administration, well-delineated as an intradural extramedullary tumor\(^1,5-7,11,13,15\) and can have a cystic component\(^6\). These findings are often first diagnosed as meningiomas or neurinomas. Therefore, although they are rare, the possibility of IDEM spinal ependymoma should be considered in the diagnosis of IDEM spinal tumors. In our case, the pre-operative diagnosis was also a meningioma versus schwannoma.

The optimal therapy for spinal IDEM ependymomas is complete resection. Macroscopically the lesion is encapsulated without apparent attachment to the central nervous system and complete removal is possible in most cases\(^1,10\) and allows preservation of normal neural tissues, although the lesions are connected to the pia of the spinal cord by microvascular attachment or to the cord by only a thin stalk in some cases\(^5\). Small tumors\(^6\) can be removed en bloc, but larger tumors\(^5,9\) may require intracapsular decompression. This can be performed safely with ultrasonic aspirator. Dense attachments may only allow a piecemeal removal of tumor to minimize the risk of significant neurological deficit. In the present case, the lesion was also encapsulated without infiltration of the spinal cord; however some parts of the tumor capsule were adherent to the spinal cord and roots. Thus, we left the adherent tumor capsule in place to minimize the risk of neurological sequelae. Most of spinal IDEM ependymomas are described as a benign lesion\(^5\), but some cases of recurrence, anaplastic transformation, and metastasis have been described in recent reports\(^1,2,6,10,11\). Although the efficacy of post-operative radiotherapy was controversial, post-operative radiotherapy was performed in our patient. The patients showed an excellent clinical recovery and no recurrence after 5 years of follow-up.

**CONCLUSION**

We report a rare case of IDEM ependymoma of cervical spine with a review of the literature.

• Acknowledgements

This work was supported for two years by Pusan National University Research Grant.

**Fig. 4.** Post-operative gadolinium-enhanced axial (A) and sagittal magnetic resonance image (B) show no definite abnormally enhanced lesion in the cervical spinal cord.

**References**

1. Benzagmout M, Boujraf S, Ouali N, Chbani L, Amarti A, Chakour K, et al.: Intradural extramedullary ependymoma: is there constantly a hormonal relationship? *Surg Neurol* 70: 536-538; discussion 538, 2008
2. Cerese A, Venturi C, Oliveri G, De Falco D, Miracco C: Intradural extramedullary spinal anaplastic ependymoma. Case illustration. *J Neurosurg Spine* 5: 476, 2006
3. Cheng CH, Lee TC, Huang HY, Lui CC: Extradural thoracic myxopapillary ependymoma—a case report of a rare tumour. *Ann Acad Med Singapore* 25: 869-872, 1996
4. Cooper JS, Craig WM, Kernohan JW: Tumors of the spinal cord: primary extradural gliomas. *Surg Gynecol Obstet* 92: 183-190, 1951
5. Dufai H, Gazzar M, Kujas M, Fohanno D: Primary intradural extramedullary ependymoma: case report and review of the literature. *Spine (Phila Pa 1976)* 25: 1993-1995, 2000
6. Graça J, Gilhaut N, D’Haeene N, Brotchi J, Salmon I, Baleaurox D: Cystic extradural ependymoma. *AJNR Am J Neuroradiol* 27: 818-821, 2006
7. Katoh S, Ikada T, Inoue M, Takahashi M: Intradural extramedullary ependymoma. A case report. *Spine (Phila Pa 1976)* 20: 2036-2038, 1995
8. Kim SY, Kim SW: Primary intradural extramedullary myxopapillary ependymoma. *J Korean Neurosurg Soc* 39: 382-384, 2006
9. Payer M, Yonekawa Y, Imhof HG: Solitary thoracic intradural extramedullary ependymoma. *J Clin Neurosci* 6: 344-345, 1999
10. Robles SG, Saldana C, Boto GR, Martinez A, Zamarron AP, Jorquera M, et al.: Intradural extramedullary spinal ependymoma: a benign pathology? *Spine (Phila Pa 1976)* 30: E521-E524, 2005
11. Schuurmans M, Vanneste JA, Verstegen M, van Furth WR: Spinal extramedullary anaplastic ependymoma with spinal and intracranial metastases. *J Neurooncol* 79: 57-59, 2006
12. Schwartz TH, McCormick PC: Spinal cord tumors in adults in Winn HR (ed): *Neurological surgery*, ed 5. Philadelphia: WB Saunders, 2004, Vol 4, pp 4817-4834
13. Wagle WA, Jaufman B, Mincy JE: Intradural extramedullary ependymoma: MR-pathologic correlation. *J Comput Assist Tomogr* 12: 705-707, 1988
14. Wahab SH, Simpson JR, Michalski JM, Mansur DB: Long term outcome with post-operative radiation therapy for spinal canal ependymoma. *J Neurooncol* 83: 85-89, 2007
15. Wolfit CE, Azzarelli B, Shah MV: Primary extramedullary ependymoma of the thoracic spine. Case illustration. *J Neurosurg* 87: 643, 1997