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Case Report

Intraventricular Atypical Meningiomas

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A rare case of intraventricular meningioma that arose in the atrium of the left lateral ventricle was identified in a 51-year-old woman. Gross total removal was performed by transcortical approach. Histopathological findings showed meningothelial meningioma with a focal atypical area which had 8% of Ki-67 labeling index (LI). A large recurrence extending into the ipsilateral quadrigeminal cistern and opposite medial occipital lobe developed approximately 41 months after the first operation. The specimens obtained from the second resection showed atypical meningioma with 20% of Ki-67 LI but there were no anaplastic area. The patient underwent fractionated stereotactic radiotherapy. However, multiple local distant metastases were found in the occipital and cerebellar cortex suggesting cerebrospinal fluid dissemination apparently 24 months after the second operation. This report presents chronological progression of a rare intraventricular atypical meningioma with more aggressive transformation.

Key Words : Atypical · Intraventricular · Meningioma · Transformation.

INTRODUCTION

Intraventricular meningiomas are rare tumor, comprising 0.5-3.7% of intracranial meningiomas in most series[2,11,14,16,18]. Characteristically, they do not have dural attachment unlike usual meningiomas. Most intraventricular meningiomas arise in the lateral ventricle, especially in the trigone[2,11,14,16]. These lesions usually show benign clinical course and indolent biological behavior. Non-benign meningiomas with intraventricular location and their clinical courses have been reported to be very few[4,6,8,9,11,12,23]. Herein, we report a rare case of intraventricular atypical transformed meningioma with relevant literature review.

CASE REPORT

A 51-year-old woman presented with right hemiparesis and gait disturbance. Magnetic resonance imaging (MRI) demonstrated a mass measuring about 5×4×4 cm in the trigone of the left lateral ventricle. The tumor showed iso-signal intensity on T1 weighted image (WI), high-signal intensity on T2WI, and homogeneous strong enhancement. Focal dilatation of the left lateral ventricle located posterior to the tumor was also noted (Fig. 1A, B, C). Gross total excision was performed through the transcortical approach. The tumor was attached to the choroid plexus but the adjacent brain parenchymal invasion was not apparent. The patient underwent craniotomy for postoperative epidural hematoma but recovered without further neurological deficits. Most specimens showed typical histopathological features of benign meningothelial meningioma. There was neither nuclear atypism nor pleomorphism and mitotic figures were less than 2 per 10 high power fields (HPF) (Fig. 2A). There were no foci of necrosis but a small focal area of atypical form with 8% of Ki-67 labeling index (LI) showing increased cellularity, small cells with a high nuclear : cytoplasmic ratio, and sheet-like growth was noted (Fig. 2B). The patient visited for the progression of hemiparesis 41 months after the first operation. MRI showed a large recurrence which extended into the ipsilateral quadrigeminal cistern and contralateral occipital lobe over the falx, and compressed splenium, thalamus (Fig. 1D). Subtotal resection was done due to tumor encasement of important venous structures. The specimens obtained from the second operation showed atypical meningioma with increased Ki-67 LI (20%) and frequent mitotic figures (Fig. 2C). An immunohistochemical study confirmed that both primary and recurred tumors were almost identical, except for the markedly increased Ki-67 LI. The patient underwent fractionated stereotactic radiotherapy (FSRT). The recurred atypical tumor and peritumoral edema were reduced gradually on follow-up enhanced computed tomography (CT). Twenty-four months after the second operation, newly developed two distant metastatic lesions in the left occipital and left cerebellar cortical area and progression of a small mass attached to the falx were demonstrated on MRI (Fig. 1E, F, G). They were considered as cerebrospinal fluid (CSF) dissemination. The patient underwent stereotactic radiosurgery

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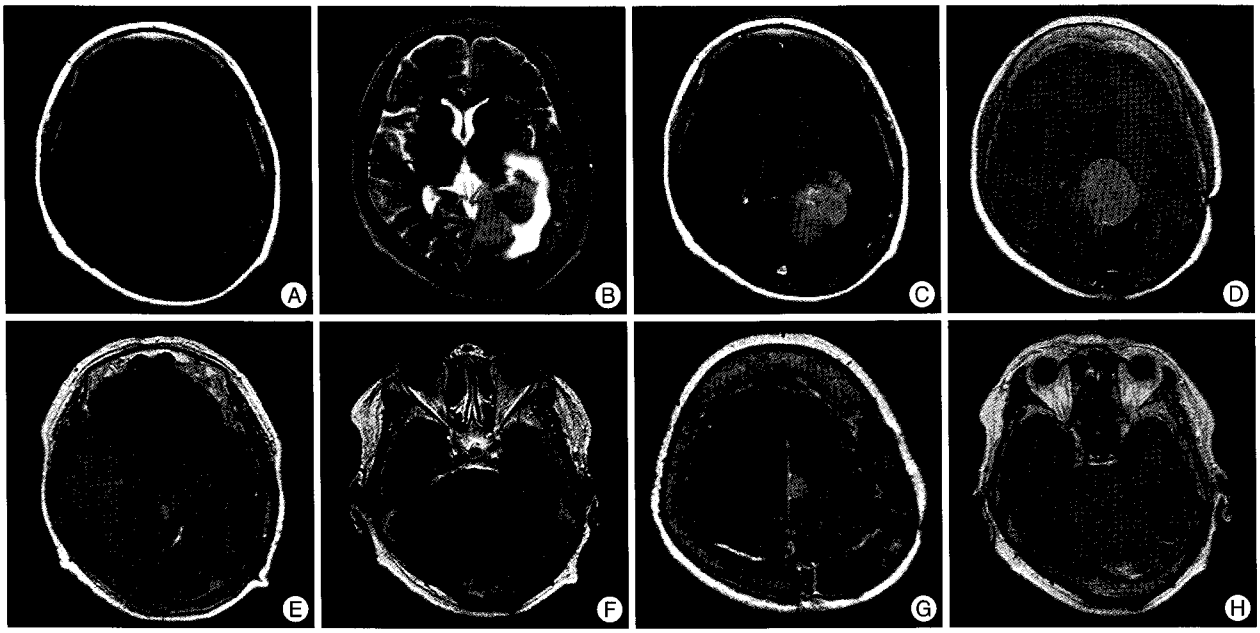

Fig. 1. MRI is showing a large mass in the trigone of the left lateral ventricle. The tumor shows iso-signal intensity on T1WI (A), high-signal intensity on T2WI (B), and homogeneous strong enhancement (C). Enhanced T1WI at the recurrence is showing that the tumor is extending into the ipsilateral quadrigeminal cistern, contralateral occipital lobe and is compressing splenium, thalamus (D). The new local distant metastatic lesions in the left occipital (E) and left cerebellar cortical area (F) and progression of a small mass attached to the falx (G) are identified on enhanced T1WI performed about 24 months after the recurrence. All the lesions are increasing in size gradually and a new enhancing lesion is found in the fourth ventricle on enhanced T1WI (H). MRI : magnetic resonance imaging, WI : weighted image.

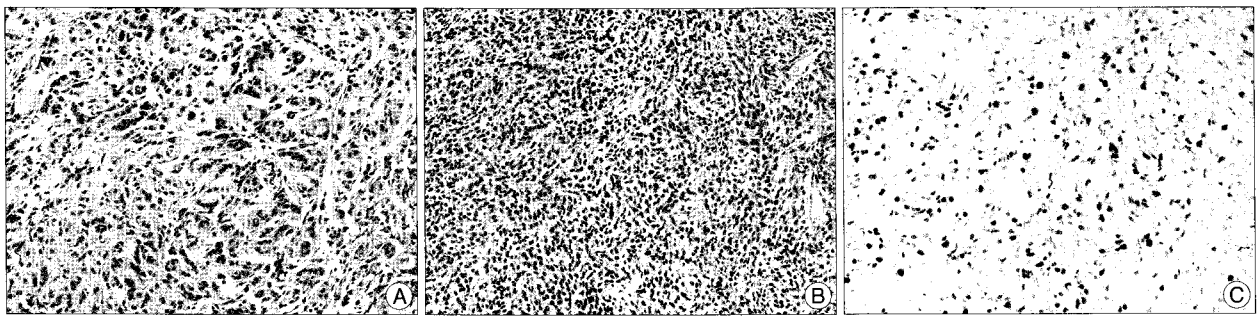

Fig. 2. Pathological findings at the first operation are showing that most area is typical benign meningothelial meningioma. There are neither nuclear atypism nor pleomorphism and mitotic figures are less than 2 per 10 (HPF) (A). However, a small focal area with atypical form showing increased cellularity, small cells with a high nuclear : cytoplasmic ratio, and sheet-like growth and 8% of Ki-67 LI are noted (B). At the time of recurrence, Ki-67 LI increased to 20% (C). HPF : high power fields, LI : labeling index.

(SRS) targeting 3 lesions. However, all the lesions increased in size gradually and a new enhancing lesion was found in the fourth ventricle (Fig. 1H).

DISCUSSION

Meningiomas arising in the ventricle without any dural attachment are rare in adult[2,11,14,16,18]. They can arise from the tela choroidea and the choroid plexus stroma[3]. Most intraventricular meningiomas are low grade, therefore follow a benign clinical course[10,14,16]. Of all meningiomas, atypical and malignant meningiomas account for 4.7-19.8% and 1-7.2% respectively[1,7,17] and intraventricular meningiomas comprise 0.5-3.7% of intracranial meningiomas[2,11,14,16,18]. Therefore, intravenricular non-benign meningiomas are considered to be less than 1% of intracranial meningiomas[2,11,14]. In addition, the incidence of non-benign meningiomas is known to be higher in younger patients[15]. To the best of our knowledge, the clinocopathological findings of intraventricular atypical meningiomas have not been described although those of malignant intraventricular meningiomas have been reported[9]. We summarized the clinicopathological findings of 4 atypical intraventricular meningiomas including our case in Table 1. In that, several scattered cases with insufficient clinicopathological descriptions were excluded[2,4,6,8,9,11,14,24]. All the cases seemed to be atypical de novo meningiomas and transformed into the anaplastic form eventually along with extraneural metastasis and/or CSF dissemination. In the transformation of meningiomas, the LOH at 22q is considered to be the earliest initiating event[17]. Secondary abnormalities involved in meningioma progression from the benign to the atypical state

Table 1. Clinicopathological findings of atypical intraventricular meningiomas

No.	Author	Sex/Age	Location	Initial histology	Time to recur	Time to transform		Histological evolution	Metastasis	Time to metastases	Survival	Treatment
						I → II	II → III					
1	Darwish et al.	F/53	Trigone	II scattered mitosis focal necrosis	4 mon (atypical)		8 mon	III mitosis up to 32/10HPF increased necrosis Ki-67 LI 20%	Local distant, spine, galea	8 mon	At least 8 mon	RT after metastasis
2	Eom et al.	F/42	Trigone	II mitosis 6-7/10HPF Ki-67 LI 7%	38 mon		38 mon	III mitosis >20/10HPF increased necrosis Ki-67 LI 25%	Spine	48 mon since II, 10 mon since III	48 mon since II, 10 mon since III	RT
3	Garcia-Conde et al.	M/44	Trigone	II mitosis <10/10HPF small necrosis Ki-67 LI 10%	2 mon		2 mon	III mitosis 15-20/10HPF wide necrosis Ki-67 LI 60%	Liver	4 mon since II, 2 mon since III	7 mon since II, 5 mon since III	RT
4	Present case	F/51	Trigone	II mitosis 2/10HPF no necrosis Ki-67 LI 8%	41 mon		65 mon (?)	II (recurrent) mitosis 3/10HPF Ki-67 LI 20%	Local distant (cerebellum, occipital)	65 mon since II	At least 74 mon	FSRT on recurrence

no. : number, II, III : WHO grade, HPF : high power field, mon. : months, RT : radiation therapy, FSRT : fractionated stereotactic radiation therapy

are associated with the allelic loss of 1p, 6q, 10q, 14q, and 18q and gains of 1q, 9q, 12q, 15q, 17q, and 20q. Furthermore, progression from the atypical to the anaplastic state is associated with the loss of 6q, 9p, 10q, and 14q, and amplification of 17q; mutations in TP53 and PTEN; and deletions in CDKN2A[13]. Therefore, it has been elucidated that the progression from ordinary, through atypical, to anaplastic meningioma is a process in which many genetic aberrations are accumulated successively[15]. CT and MRI findings of intraventricular meningiomas are relatively nonspecific but irregular lobulated shape and intratumoral necrosis are frequently seen in the atypical or malignant forms[11]. Heterogeneous signal intensities both on T1 and T2 WI and the heterogeneous enhancing pattern should raise the suspicion of a malignant meningioma[3]. In our case, MRI showed the similar findings between primary and recurrent tumors but slightly higher signal intensity on T2WI in recurrent tumor. More irregular shape and more heterogeneous signal pattern were seen in MRI performed at the time of local distant metastases suggesting of anaplastic transformation. According to the reports, the overall rate of progression from benign to higher-grade tumor ranges from 0.16 to 2% and the risk of progression from atypical to malignant form ranges from 26 to 33%[1,20,22]. In the intraventricular malignant meningiomas, the cases of anaplastic intraventricular meningiomas transformed from benign or atypical form has increased[8,9,23]. Therefore, it suggests that anaplastic transformation may be frequent or usual once a benign form progress to an atypical form. And also, it seems that even atypical or anaplastic de novo forms may be consecutively transformed from more benign form over variable period although the hypothesis of direct alternative pathway progressing from benign to anaplastic form has been postulated[12]. There is also a trend towards progressive evolution to more malignant form when recurrences follow[5]. In our review cases, progressive histopathological evolutions were also usual. The possibility of metachronous meningioma seems very low in the absence of any stigmata of neurofibromatosis and distant mass with different radiological evolution. One case (No. 3) transformed into the anaplastic form showed an extraneural metastasis to the liver without definite CSF seeding. However, malignant intraventricular meningiomas seem to have a tendency to spread easily through the CSF[8,9]. In these cases, spinal tap may be helpful to diagnose the metastasis via CSF such as Darwish et al.[6] recommendation. Our present case (No. 4) showed local distant metastases suggesting of CSF dissemination but spinal tap was not performed. Immunohistochemical tests such as the Ki-67 proliferation index are very important. They are helpful to differentiate atypical meningiomas from anaplastic types and may also help to predict the recurrence rate[9,21]. The Ki-67 LI increased with the tumor grade significantly and supported the usefulness of the Ki-67 LI as a supplementary tool for grading[19]. Our present case showed increased Ki-67 LI (20%) at the recurrence despite same histological grade. In our review cases, it was shown that the time to recur and transform was variable but histopathological evolu-

tions and increased Ki-67 LIs were constant. It was reported that transformation period from intraventricular benign meningioma to anaplastic form were 4-5 years[9]. It is considered that transformation period from atypical meningioma to anaplastic form seems to be shorter than that from benign meningioma to atypical form compared with our review cases despite small sample size. Usually, average survival rate of atypical meningioma and malignant intraventricular meningioms are approximately 2-5 years and 9 months respectively[9,12]. In our review cases, average survival rate of intraventricular atypical meningiomas was similar with that of intracranial atypical meningiomas as approximately 34 months and longer than malignant intraventricular meningiomas.

CONCLUSION

Atypical intraventricular meningioma is rare. Further anaplastic progression over relatively short period should be anticipated. Therefore, more aggressive treatment and meticulous follow-up are required in the atypical intraventricular meningiomas.

References

1. Al-Mefty O, Kadri PA, Pravdenkova S, Sawyer JR, Stangeby C, Husain M : Malignant progression in meningioma: documentation of a series and analysis of cytogenetic findings. **J Neurosurg 101** : 210-218, 2004
2. Bertalanffy A, Roessler K, Koperek O, Gelpi E, Prayer D, Neuner M, et al. : Intraventricular meningiomas : a report of 16 cases. **Neurosurg Rev 29** : 30-35, 2006
3. Chen NF, Lin GY, Wang YC, Leu CH, Kwan PC : Intraventricular malignant meningioma : one case report. **J Clin Neurosci 10** : 616-620, 2003
4. Cleary C, Curtin D : Giant atypical intraventricular meningioma presenting with visual loss in a child. **Ir J Med Sci 179** : 617-619, 2010
5. Conrad MD, Schonauer C, Pelissou-Guyotat I, Morel C, Madarassy G, Deruty R : Recurrent lumbosacral metastases from intracranial meningioma. Report of a case and review of the literature. **Acta Neurochir (Wien) 143** : 935-937, 2001
6. Darwish B, Munro I, Boet R, Renaut P, Abdelaal AS, MacFarlane MR : Intraventricular meningioma with drop metastases and subgaleal metastatic nodule. **J Clin Neurosci 11** : 787-791, 2004
7. Enam SA, Abdulrauf S, Mehta B, Malik GM, Mahmood A : Metastasis in meningioma. **Acta Neurochir (Wien) 138** : 1172-1177; discussion 1177-1178, 1996
8. Eom KS, Kim HS, Kim TY, Kim JM : Intraventricular Malignant Meningioma with CSF-disseminated spinal metastasis : case report and literature review. **J Korean Neurosurg Soc 45** : 256-259, 2009
9. Garcia-Conde M, Roldan-Delgado H, Martel-Barth-Hansen D, Manzano-Sanz C : Anaplastic transformation of an atypical intraventricular meningioma with metastases to the liver : case report. **Neurocirugia (Astur) 20** : 541-549, 2009
10. Guidetti B, Delfini R, Gagliardi FM, Vagnozzi R : Meningiomas of the lateral ventricles. Clinical, neuroradiologic, and surgical considerations in 19 cases. **Surg Neurol 24** : 364-370, 1985
11. Kim EY, Kim ST, Kim HJ, Jeon P, Kim KH, Byun HS : Intraventricular meningiomas: radiological findings and clinical features in 12 patients. **Clin Imaging 33** : 175-180, 2009
12. Krayenbühl N, Pravdenkova S, Al-Mefty O : De novo versus transformed atypical and anaplastic meningiomas : comparisons of clinical course, cytogenetics, cytokinetics, and outcome. **Neurosurgery 61** : 495-503; discussion 503-504, 2007
13. Lee JY, Finkelstein S, Hamilton RL, Rekha R, King JT Jr, Omalu B : Loss of heterozygosity analysis of benign, atypical, and anaplastic meningiomas. **Neurosurgery 55** : 1163-1173, 2004
14. Liu M, Wei Y, Liu Y, Zhu S, Li X : Intraventricular meninigiomas : a report of 25 cases. **Neurosurg Rev 29** : 36-40, 2006
15. Mahmood A, Caccamo DV, Tomecek FJ, Malik GM : Atypical and malignant meningiomas : a clinicopathological review. **Neurosurgery 33** : 955-963, 1993
16. Menon G, Nair S, Sudhir J, Rao R, Easwer HV, Krishnakumar K : Meningiomas of the lateral ventricle - a report of 15 cases. **Br J Neurosurg 23** : 297-303, 2009
17. Modha A, Gutin PH : Diagnosis and treatment of atypical and anaplastic meningiomas: a review. **Neurosurgery 57** : 538-550; discussion 538-550, 2005
18. Nakamura M, Roser F, Bundschuh O, Vorkapic P, Samii M : Intraventricular meningiomas : a review of 16 cases with reference to the literature. **Surg Neurol 59** : 491-503; discussion 503-504, 2003
19. Nakane Y, Natsume A, Wakabayashi T, Oi S, Ito M, Inao S, et al. : Malignant transformation-related genes in meningiomas : allelic loss on 1p36 and methylation status of p73 and RASSF1A. **J Neurosurg 107** : 398-404, 2007
20. Palma L, Celli P, Franco C, Cervoni L, Cantore G : Long-term prognosis for atypical and malignant meningiomas: a study of 71 surgical cases. **J Neurosurg 86** : 793-800, 1997
21. Pramesh CS, Saklani AP, Pantvaidya GH, Heroor AA, Naresh KN, Sharma S, et al. : Benign metastasizing meningioma. **Jpn J Clin Oncol 33** : 86-88, 2003
22. Rohringer M, Sutherland GR, Louw DF, Sima AA : Incidence and clinicopathological features of meningioma. **J Neurosurg 71** : 665-672, 1989
23. Shintaku M, Hashimoto K, Okamoto S : Intraventricular meningioma with anaplastic transformation and metastasis via the cerebrospinal fluid. **Neuropathology 27** : 448-452, 2007
24. Takara K, Cho K, Mori K, Maeda M : Intraventricular, cystic, atypical meningioma. **Neurol Med Chir (Tokyo) 37** : 856-860, 1997