J Korean Radiol Soc 2003;49:155 - 164

Various Tumors in the 4th Ventricle in Adults: MRI Findings[1]

Seung Ja Kim, M.D., Kee-Hyun Chang, M.D., Keon Ha Kim, M.D., Ja Young Choi, M.D., Bae Joo Kwon, M.D., Moon Hee Han, M.D.

Primary fourth ventricular neoplasms are common in children but rare in adults, and the disease categories encountered differ according to the patient's age. This study reviewed the records of patients aged 16 years or over, who underwent magnetic resonance (MR) imaging and were found to have fourth ventricular lesions. Most patients then underwent surgical resection, leading to specific pathologic diagnosis. The various fourth ventricular tumors encountered were ependymoma ($n=8$), subependymoma ($n=1$), choroid plexus papilloma ($n=3$), astrocytoma ($n=3$), medulloblastoma ($n=1$), lymphoma ($n=2$), epidermoid cyst ($n=2$), meningioma ($n=1$), melanoma ($n=1$), cavernous hemangioma ($n=1$) and metastasis ($n=1$).

We describe the various tumors located mainly in the fourth ventricle and review their clinical presentation and the radiological findings, the majority of which were nonspecific. In some cases, however, specific signal intensity or the growth pattern of the tumors was useful for differential diagnosis.

Index words : Brain neoplasms
Ventricles
Magnetic resonance (MR)

Primary fourth ventricular tumors are common in children but rare in adults. In addition, the disease categories encountered differ according to the patient's age; in adults, the most common fourth ventricular neoplasm is metastasis (1). The most frequent primary intraventricular tumor of the posterior fossa is the ependymoma; other tumors include choroid plexus papillomas and an occasional meningioma (2). Subependymomas can occur in the inferior fourth ventricle near the obex and are usually found incidentally in older patients (1).

[1]Department of Radiology, Seoul National University College of Medicine and the Institute of Radiation Medicine, SNUMRC
Received April 16, 2003 ; Accepted July 11, 2003
Address reprint requests to : Kee-Hyun Chang, M.D., Department of Radiology, Seoul National University Hospital, 28, Yongon-dong, Chongno-gu, Seoul 110-744, Korea.
Tel. 82-2-760-2584 Fax. 82-2-743-6385
E-mail: changkh@radcom.snu.ac.kr

This paper describes the various tumors located mainly in the fourth ventricle in adults, and reviews their clinical presentation and the radiological findings.

Ependymoma

Ependymomas account for 2 - 8% of all primary intracranial brain tumors, and are four to six times more common in children than in adults. MRI differentiates ependymomas from other gliomas on the basis of their location and morphology, but the signal intensity (SI) or enhancement patterns seen at MRI are similar. The classic appearance of a posterior fossa ependymoma is a somewhat lobulated soft-tissue mass that appears to form a cast or mold of the fourth ventricle and extrudes through its outlet foramina into the adjacent subarachnoid cisterns (3). An ependymoma can arise anywhere

in the ventricular system, but the fourth ventricle, particularly the caudal part of its floor, is the most common site, followed in order of frequency by the lateral ventricle, the aqueduct, the third ventricle, the spinal cord, the filum terminale and the cerebral hemisphere (4). This study reviewed eight cases of fourth ventricular ependymoma, the solid components of which were a low or iso SI compared to the brain at T1-weighted imaging and a high SI at T2-weighted imaging. In six of the eight cases, the mass extended to the level of the foramen magnum, and in three cases, intratumoral hemorrhage was detected (Figs. 1, 2).

Subependymoma

A subependymoma is a rare benign tumor of the central nervous system (CNS), in which age at presentation, histology, and clinical course are distinct. Subependymomas occur in middle-aged and elderly adults, most often arising from the lower medulla and projecting into the caudal fourth ventricle (3). Chiechi et al. (5) reported nine fourth ventricular subependymomas, eight of which extended into the foramina of Luschka/ Magendie. Subependymomas tend to be well-defined intraventricular masses that enhance after contrast injection and have a high SI at T2-weighted imaging (2). We present one case in which a subependymoma showed good enhancement at enhanced T1-weighted imaging (Fig. 3).

Papilloma of the choroid plexus

Infratentorial choroid plexus papillomas typically arise from the roof of the fourth ventricle and expand into the ventricular cavity. A few, however, arise within the foramen of Luschka and present as cerebellopontine angle masses. Fourth ventricular choroid plexus papillomas account for less than 1% of all primary intracranial tumors, and in contrast to lateral ventricular choroid plexus papillomas, generally occur in adults. Hydrocephalus is common, and can result from obstruction of the cerbrospinal fluid (CSF) pathways, excessive CSF production, or both. At MRI, the lesions appear to show an iso to mildly low SI relative to adjacent cerebellar and brainstem parenchyma on T1-weighted images, but significantly higher intensity on T2-weighted images. Serpentine, branching, or tubular signal void structures reflect the high vascularity of these tumors (6). We experienced three cases of choroid plexus papilloma in adults whose mean age was 31 years. Mass size ranged from 3 to 4 cm. In all three cases, hydrocephalus was present and SI was heterogeneously low at T1-weighted

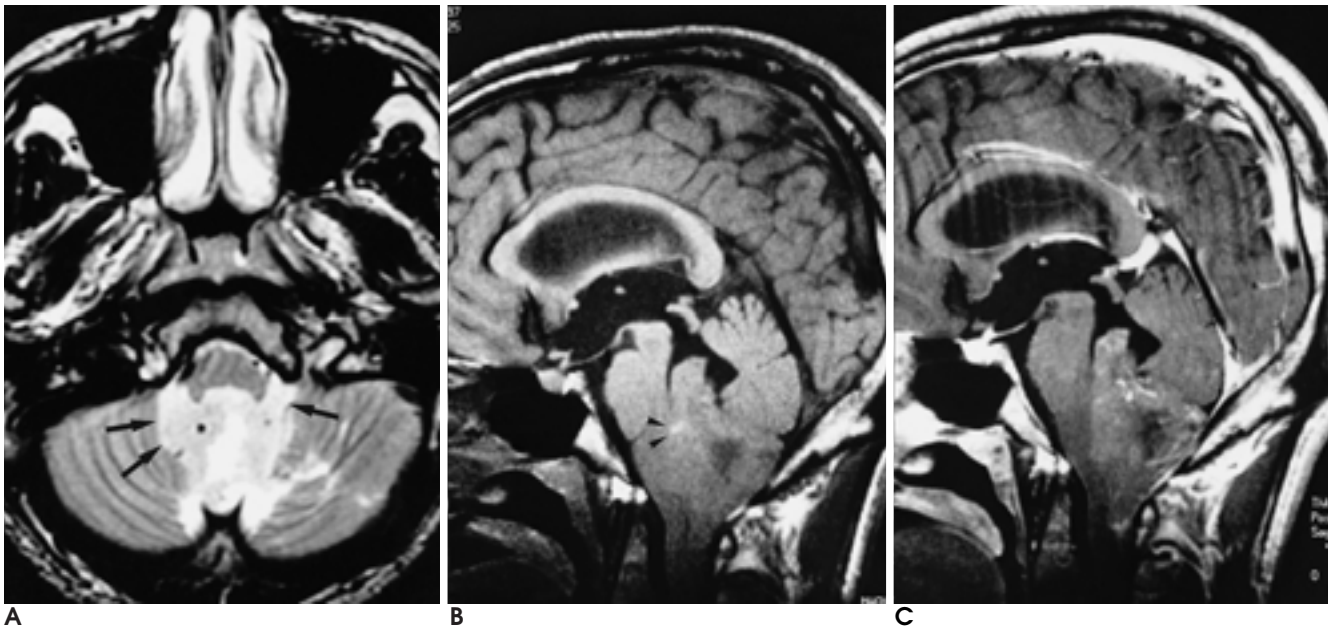


Fig. 1. Fourth venrticle ependymoma in a 27-year-old man.
A. There is an intraventricular mass in the fourth ventricle, which extends through bilateral foramen Luschka (arrows).
B. The tumor has a mainly iso SI in relation to the white matter at T1-weighted sagittal imaging and the mass has a hemorrhagic portion within it (arrowheads).
C. The mass is located in the fourth ventricle and extends to the level of the cervical cord. The mass shows heterogeneous enhancement at Gd-DTPA-enhanced T1-weighted sagittal imaging. The mass caused mild obstructive hydrocephalus.

imaging and heterogeneously high at T2-weighted imaging. In two of the three cases, the lesions extended to the lateral recess and foramen magnum. A signal void structure was detected within the mass in one case (Fig. 4).

Astrocytoma

Approximately half of all primary brain tumors are

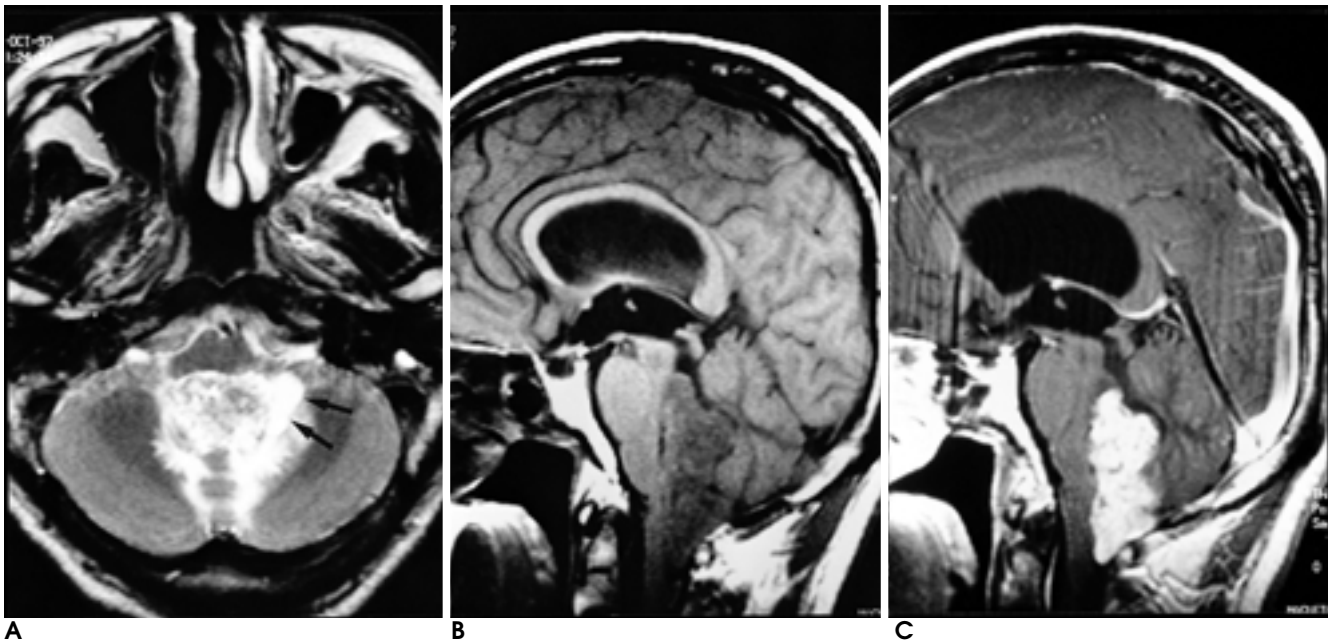


Fig. 2. Fourth ventricle ependymoma in a 57-year-old man.
A. At T2-weighted axial imaging, the fourth ventricular mass shows heterogeneous high SI. The peritumoral cyst (arrows) can be seen in the left lateral aspect of the mass.
B, C. The mass shows heterogeneous low SI at T1-weighted sagittal imaging (**B**) and relatively good homogeneous enhancement at Gd-DTPA-enhanced T1-weighted imaging (**C**). The mass also extends to the level of the cervical cord.

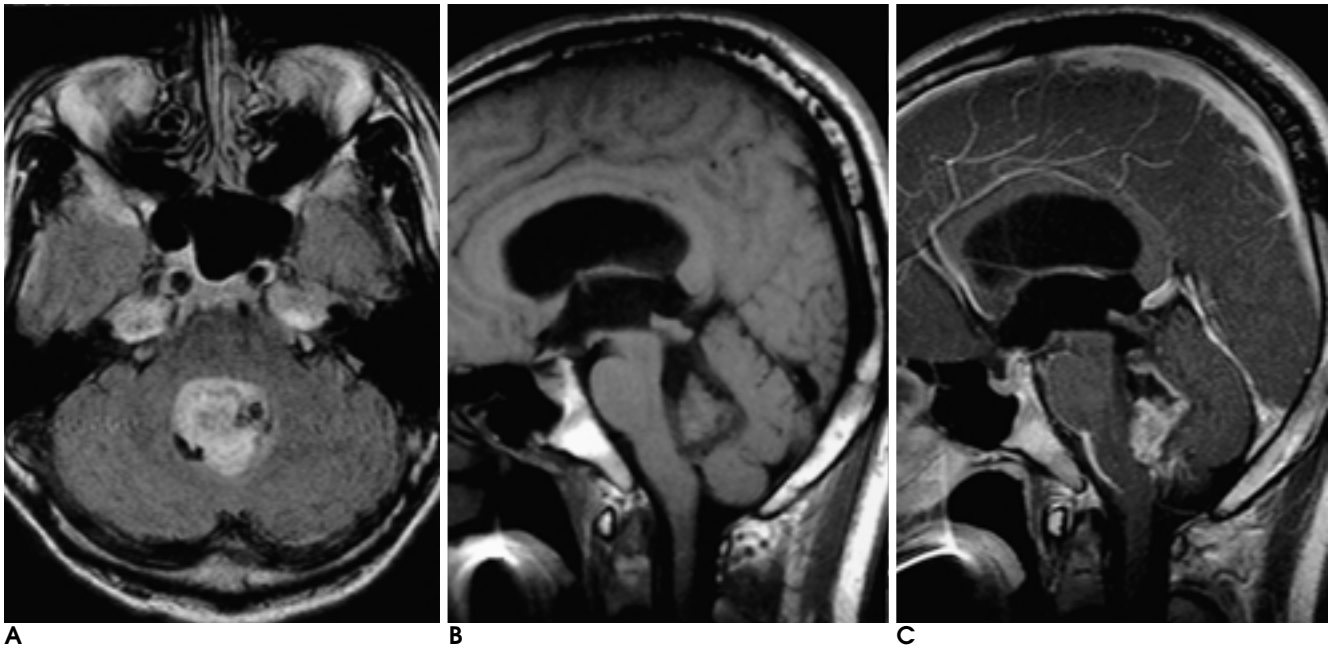


Fig. 3. Subependymoma of the fourth ventricle in a 28-year-old man.
A. There is a well-defined heterogeneous high SI mass in the fourth ventricle at T2-weighted axial imaging.
B, C. The mass shows mixed iso and low SI at T1-weighted sagittal imaging (**B**) and good heterogeneous enhancement at enhanced T1-weighted imaging (**C**).

glial cell neoplasms and more than three quarters of all gliomas are astrocytomas. Astrocytomas are a histologically heterogeneous group of primary brain tumors that have been both graded and classified, and can be subdivided and studied according to histological type, patient age, and geographic location. The generally accepted astrocytoma subtypes are fibrillary (subdivided into low-grade astrocytoma, anaplastic astrocytoma, and glioblastoma multiforme), pilocytic, and subependymal giant-cell astrocytomas, as well as pleomorphic xanthoastrocytoma. Most pilocytic astrocytomas are located around the third and fourth ventricles, whereas anaplastic astrocytomas and glioblastomas are primarily hemispheric neoplasms, which rarely occur in the posterior fossa (3). The MR imaging findings of astrocytomas vary according to their grade. We encountered an anaplastic astrocytoma, a low-grade astrocytoma and a pilocytic astrocytoma in the fourth ventricle. The low-grade fibrillary astrocytoma showed heterogeneous, low SI at T1-weighted imaging, and a heterogeneous, high SI at T2-weighted imaging. The mass had a cystic portion, and heterogeneous strong enhancement was observed after

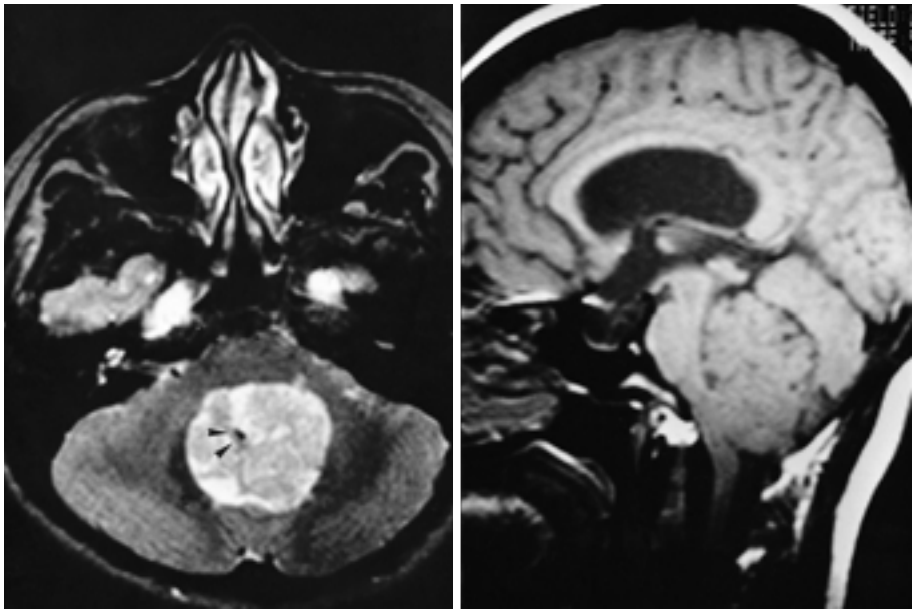


Fig. 4. Choroid plexus papilloma of the fourth ventricle in a 33-year-old woman.
A. At T2-weighted axial imaging, the mass shows heterogeneous high SI and a signal void vascular structure (arrowheads) is seen within the mass.
B. The lobulating contoured mass shows iso or slightly low SI at T1-weighted sagittal imaging and the mass extends to the level of the cervicomedullary junction.

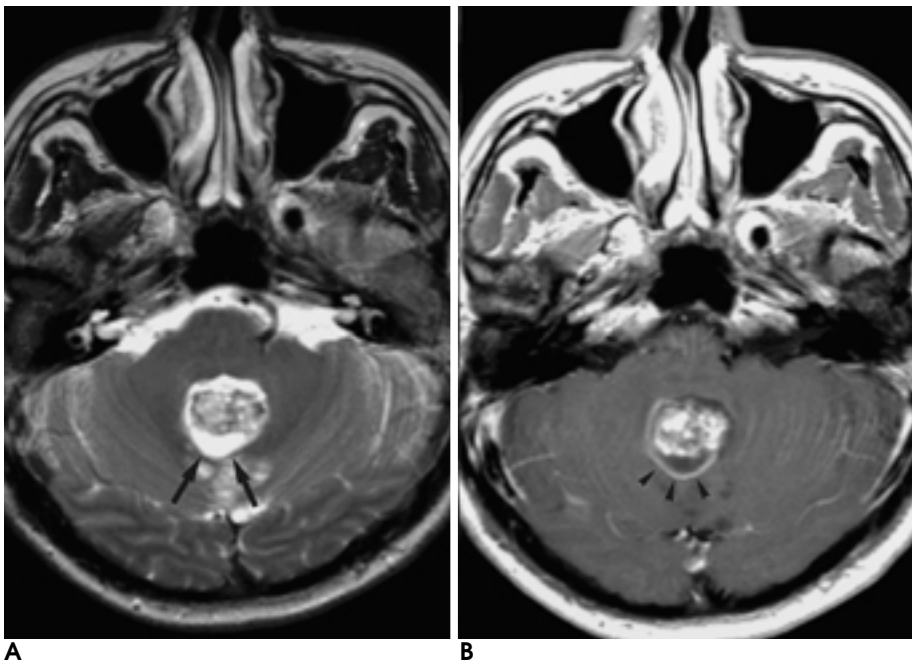


Fig. 5. Low grade astrocytoma of the fourth ventricle in a 51-year-old man.
A. At T2-weighted axial imaging, the mass shows heterogeneous SI and has cystic portion in the periphery (arrows).
B. The mass shows heterogeneous enhancement with linear enhancement along cystic portion (arrowheads) at Gd-DTPA-enhanced imaging (**B**).

contrast enhancement (Fig. 5).

Medulloblastoma

Medulloblastomas are believed to be primitive neuroectodermal tumors, and are the most common variant of this tumor category to occur in the CNS (6). They arise primarily, but not exclusively, during childhood, with almost three quarters of cases occurring before the age of 15. Medullobalstomas occur in two posterior fossa locations; the most common site, observed in 75% of cases, is the vermis, near the roof of the fourth ventricle, and tumors occurring there are midline lesions that bulge anteriorly into this ventricle and posteriorly into the cisterna magna. A less common location, occurring more commonly in older children and adults, is the cerebellum (7). Hemorrhage, cysts, and calcification are uncommon features, but in larger lesions central necrosis tends to occur. The cranial and spinal seeding of medulloblastomas via the CSF is common and often extensive (6).

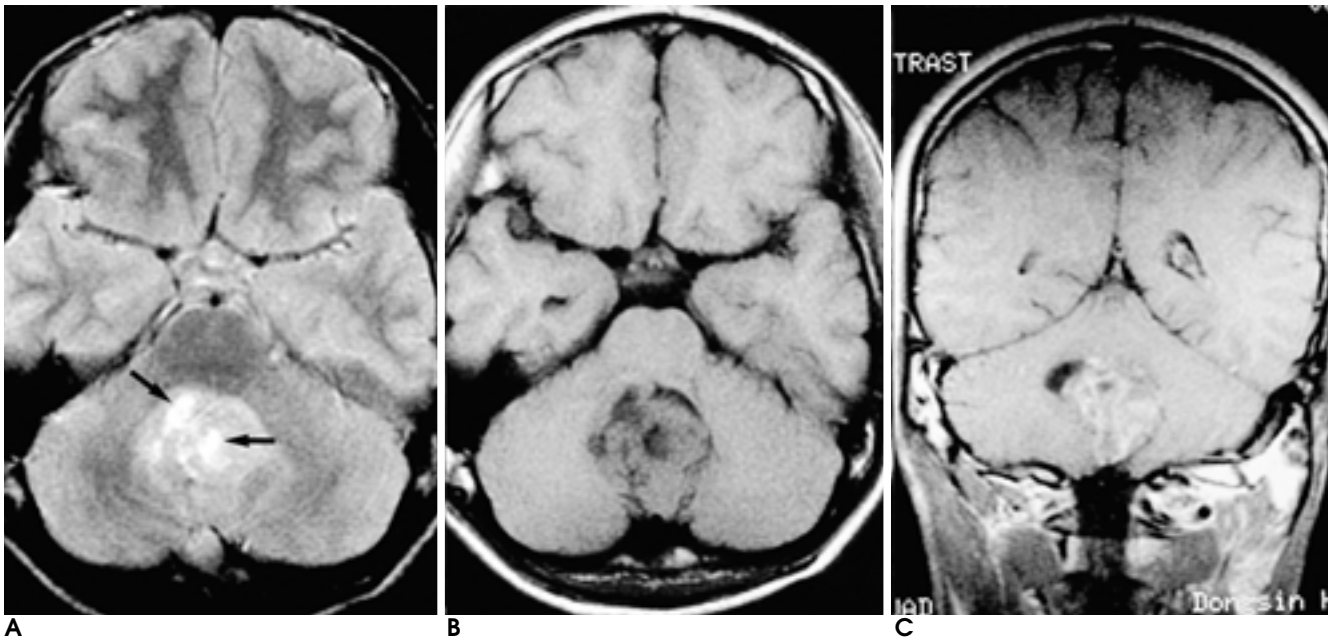
We encountered a case of medulloblastoma in a 17-year-old male (Fig. 6). T1-weighted axial imaging showed that the lesion was composed of an iso SI portion and multiple low SI areas, suggesting cystic change, and at Gd-DTPA-enhanced T1-weighted coronal imaging, good heterogeneous enhancement was apparent.

Lymphoma

A primary lymphoma of the CNS can occurs in two different settings: immunologically normal patients, and immunoincompetent patients. These tumors are found in patients of all ages: the average age in immunologically normal patients is 60 years, and in AIDS-related lymphoma sufferers, is 33 years. Primary CNS lymphomas account for approximately 1 to 2% of all intracranial neoplasms, and for approximately 1% of all primary non-Hodgkin lymphomas. Approximately 20 to 40% of such lesions are multiple. Most focal primary CNS lymphomas show iso to slightly low SI compared to gray matter at T1-weighted imaging, and iso to high SI at T2-weighted imaging. Three quarters of all primary CNS lymphomas in immunologically normal patients enhance strongly and homogeneously (7). We encountered two cases of fourth ventricular lymphoma with typical homogeneous SI and homogeneous enhancement in immunologically normal patients; in one, a second lesion was present in the pineal gland (Fig. 7).

Epidermoid cyst

Congenital epidermoid cysts probably arise from ecto-



A B C

Fig. 6. Medulloblastoma located in the fourth ventricle, vermis and right cerebellar hemisphere in a 17-year-old male.
A. At T2-weighted axial imaging at the level of pons, the mass is composed of an iso SI in relation to the gray matter and multiple high SI areas suggestive of cystic changes (arrows).
B. At T1-weighted axial imaging, the iso SI portion remains unchanged. Multiple low SI areas can be seen.
C. The mass shows good heterogeneous enhancement at Gd-DTPA-enhanced T1-weighted coronal imaging.

dermal epithelial elements included at the time of neural tube closure or during the formation of secondary cerebral vesicles. Epidermoid cysts represent 0.2 to 1% of all primary intracranial tumors and typically occur between the ages of 20 and 60. Intra-axial epidermoid tumors are, however, uncommon (8). The most frequent infratentorial location of epidermoid cysts is the cerebellopontine angle; lesions within the fourth ventri-

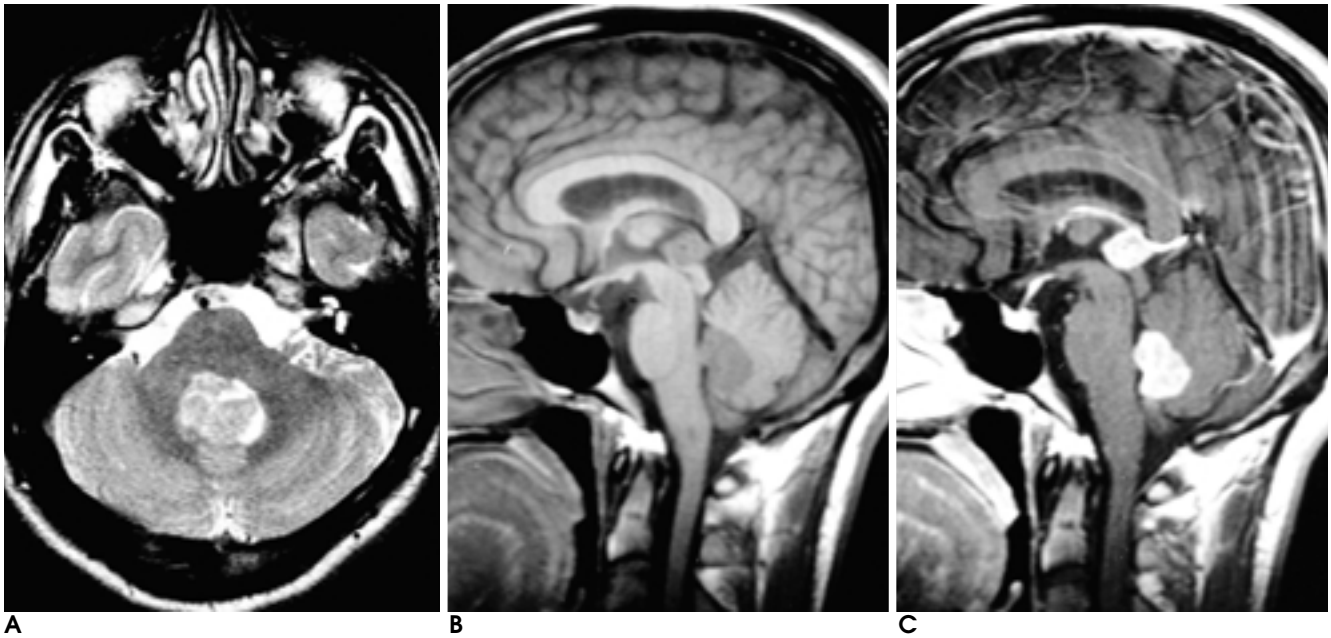


Fig. 7. Primary CNS lymphoma of the fourth ventricle and pineal area in a 23-year-old man.
A. At T2-weighted axial imaging, the mass shows homogeneous iso SI in relation to the gray matter and have lobulating contour and discrete margin.
B. The masses in the fourth ventricle and pineal gland show homogeneous slightly low SI at T1-weighted sagittal imaging.
C. The masses show good homogenous enhancement at Gd-DTPA-enhanced T1-weighted sagittal imaging.

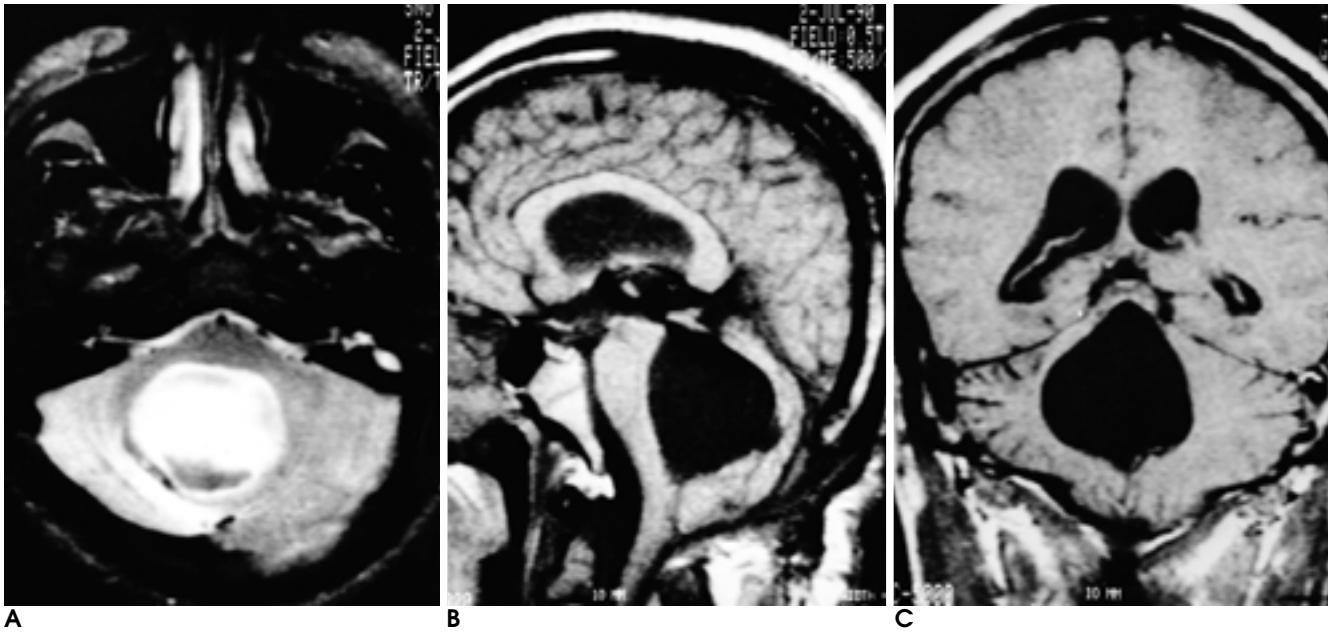


Fig. 8. Fourth ventricle epidermoid cyst in a 27-year-old man.
A. At T2-weighted axial imaging, the mass shows mainly bright SI with focal low SI in the dependent portion.
B. The cystic mass shows the same signal intensity as the CSF at T1-weighted sagittal imaging.
C. No definite enhancing portion was detected at enhanced T1-weighted coronal imaging.

cle are extremely rare (6, 9). With few exceptions, epidermoid cysts show a lower SI than adjacent brain parenchyma at T1-weighted imaging, and a high SI at T2-weighted imaging. The differential signal relative to CSF is largely related to increased protein content, which causes T1 shortening (6). We describe a case involving a 27-year-old man with a fourth ventricular epidermoid cyst showing CSF-like SI at T1-weighted imaging and no enhancement (Fig. 8).

Meningioma

Meningiomas are the most common nonglial primary brain tumor and the most common intracranial extra-axial neoplasm (7). In a larger clinical series, slightly more than 15% of meningiomas involved the posterior fossa (6). The posterior surface of the petrous temporal bone and the clivus are the most common infratentorial locations, though approximately 2% of intracranial meningiomas have no dural attachment. These tumors arise from choroid plexus stromal cells or tela choroidea, and grow as intraventricular masses (7). The majority of intraventricular meningiomas are found in the trigone of the lateral ventricle; meningiomas of the fourth ventricle are very rare, accounting for 5% of intraventricular meningiomas (10). Most meningiomas show an iso or

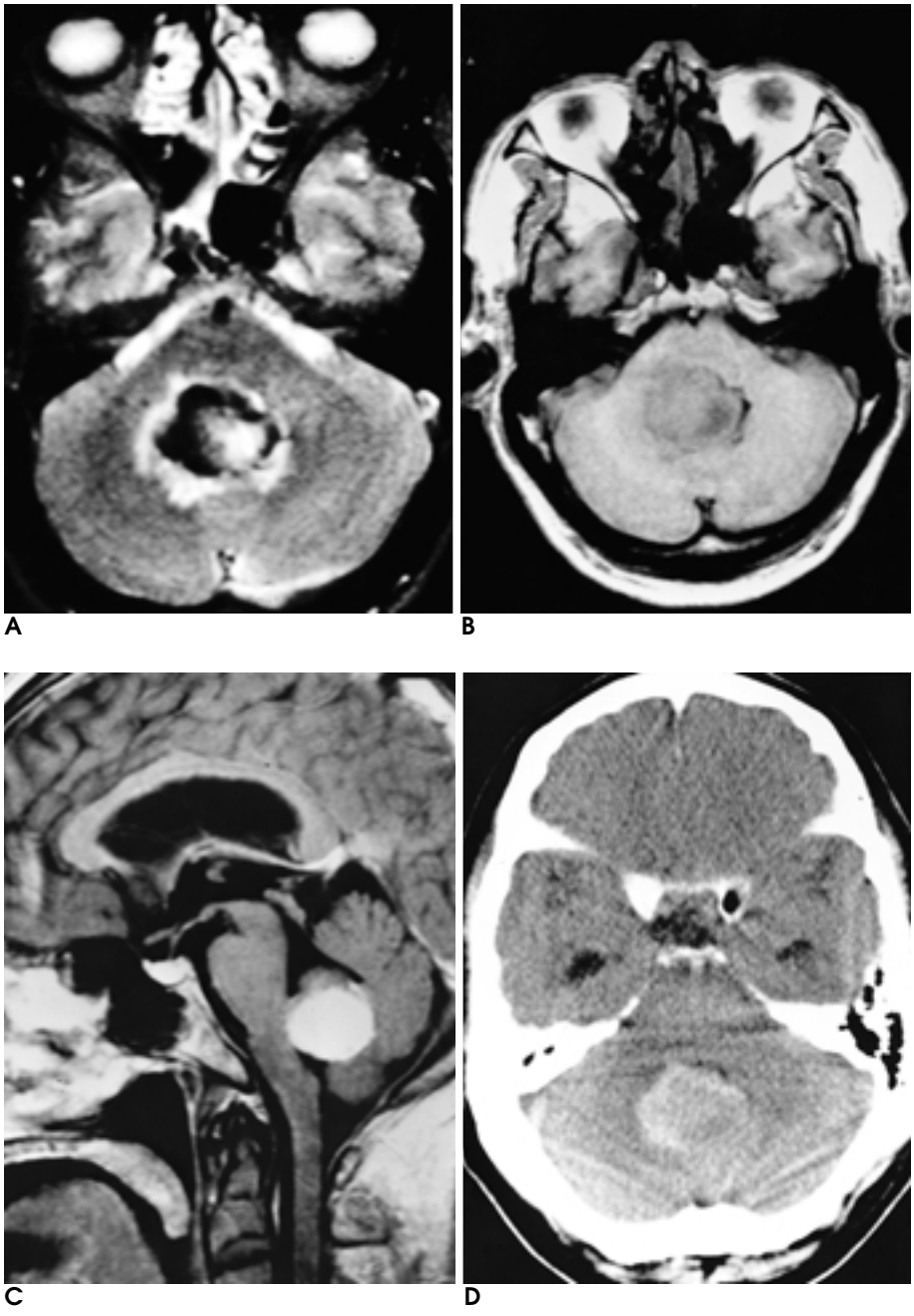


Fig. 9. Fourth ventricle meningioma in a 61-year-old man.
A. At T2-weighted axial imaging, the mass shows heterogeneous SI with dark SI portion suggestive of calcification in the periphery.
B. The mass shows mixed iso and low SI at T1-weighted axial scanning.
C. At enhanced T1-weighted sagittal imaging, the mass shows good homogeneous enhancement.
D. At precontrast CT scanning, a well-defined mass with peripheral calcification can be seen in the fourth ventricle.

slightly low SI relative to gray matter at T1-weighted imaging, though the signal observed at T2-weighted imaging can vary according to the lesion's histopathology (7). Meningiomas can be diagnosed if their contour is smooth; in such cases they enhance brightly and the fine calcification they contain is apparent (2). We present a case involving a 61-year-old man in which a fourth ventricular meningioma showed good homogeneous enhancement and peripheral calcification was suspected (Fig. 9).

Melanoma

Primary malignant melanoma most commonly originates in the skin, mucosa, and the ocular choroid; in most cases, melanomas involving the CNS have arisen through metastasis. In the CNS, particularly in the leptomeninges, melanocytes are normally found, though melanin is not usually present in the ventricular system. During brain development, melanocytes initially located within the pia might find their way into the choroid plexus, and because of the stable free radicals it contains, melanin causes T1 and T2 shortening. In the radiological literature, we found only one description of the MR imaging findings of an intraventricular melanoma. The mass was located in the left lateral ventricle and showed inhomogeneous SI, with peripheral high SI at T1-weighted imaging and relatively low SI at T2-weighted imaging (11). We present a case in which a fourth ventricular melanoma with slightly high SI at T1-weighted imaging occurred in a 36-year-old woman (Fig. 10).

Cavernous hemangioma

Cavernous hemangiomas are uncommon vascular hamartomas of the CNS, and in the ventricles they occur very rarely. In a series of intracranial cavernous hemaniomas, an intraventricular location was observed in 2.5 to 10.8% of cases. The reported characteristic MRI findings of an intraventricular cavernous hemangioma are a central area of high SI correlated with the presence of methemoglobin, together with areas of low SI apparent at T1- and T2-weighted imaging and caused by calcification and fibrosis within the lesion. A peripheral low-SI rim caused by the hemosiderin paramagnetic effect is often present, with a moderate mass effect, mild perifocal edema, and slight enhancement after gadolinium infusion (12). We present a case in which a 63-year-old woman presented with a fourth ventricular cavernous hemangioma which at T2-weighted axial imaging showed inhomogeneous high SI together with peripher-

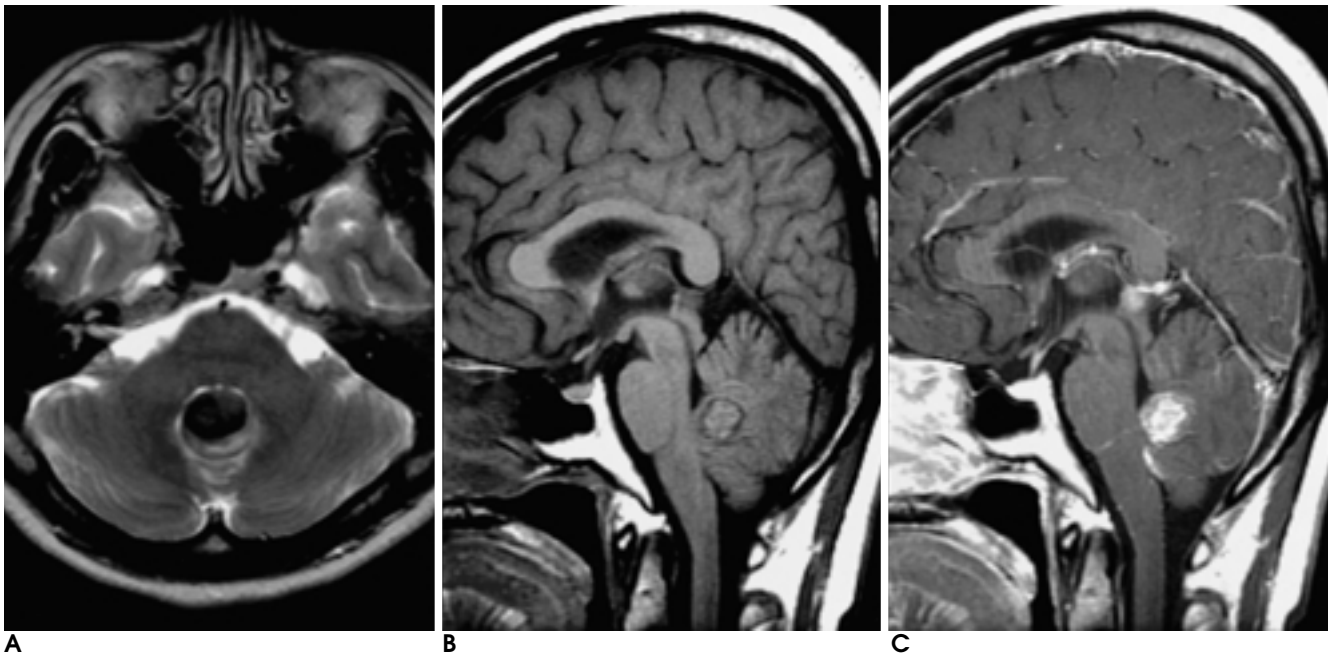


Fig. 10. Fourth ventricle melanoma in a 36-year-old woman.
A. At T2-weighted axial imaging, the fourth ventricular lesion shows heterogeneous SI and the majonty of the mass shows dark or low SI.
B. At T1-weighted sagittal imaging, the mass has a slightly high SI with a peripheral dark signal intensity rim.
C. The mass shows relatively good enhancement at enhanced T1-weighted sagittal imaging.

al dark SI, probably due to hemosiderin (Fig. 11).

Metastasis

In adults, a fourth ventricular tumor arising through metastasis is the most common type (1). A diagnosis of leptomeningeal or ventricular metastasis contributes to earlier treatment and sometimes affects decisions regarding the management of patients with intra- or extracranial malignancy. Chou et al. (13) reported that the mass or nodule present within the ventricles could be clearly identified at enhanced CT scanning, and the involved ventricles, in order of frequency, were the lateral, third, and fourth.

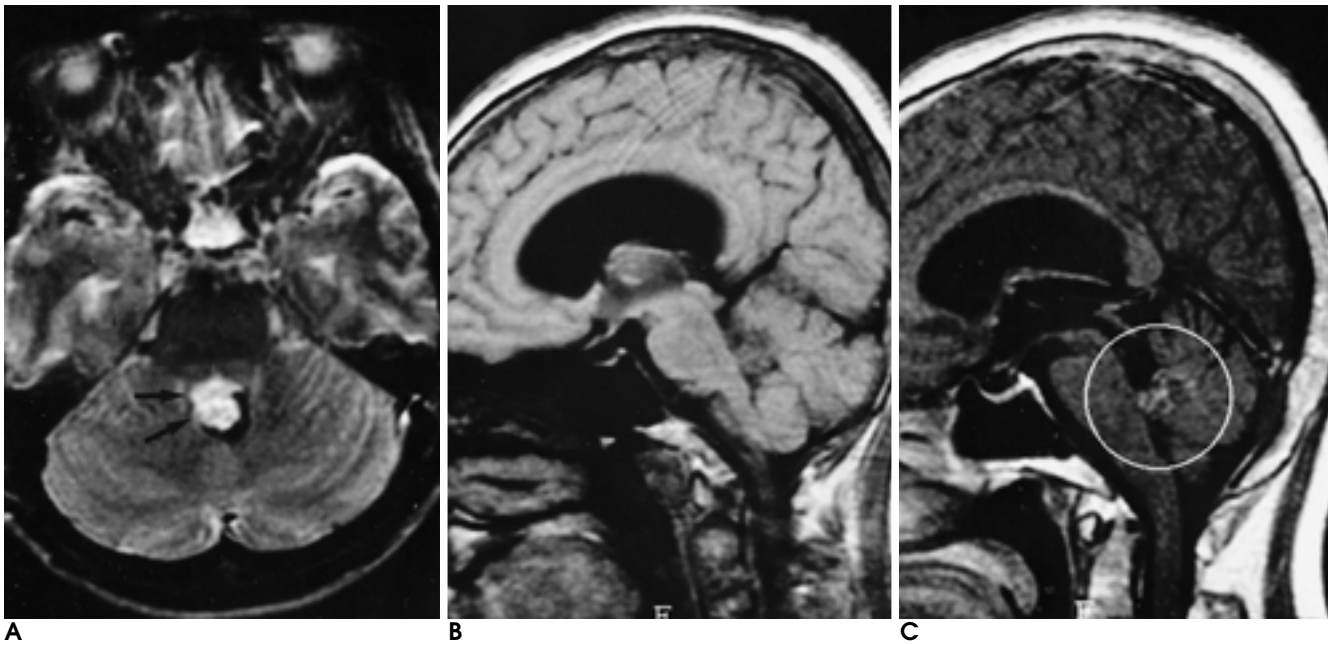


Fig. 11. Cavernous hemangioma of the fourth ventricle in a 63-year-old woman.
A. At T2-weighted axial imaging, the mass shows inhomogeneous high SI (arrows) with peripheral dark SI probably due to hemosiderin.
B. The mass shows relatively homgeneous low SI at T1-weighted sagittal imaging.
C. At enhanced T1-weighted imaging, the mass has heterogeneous enhancement.

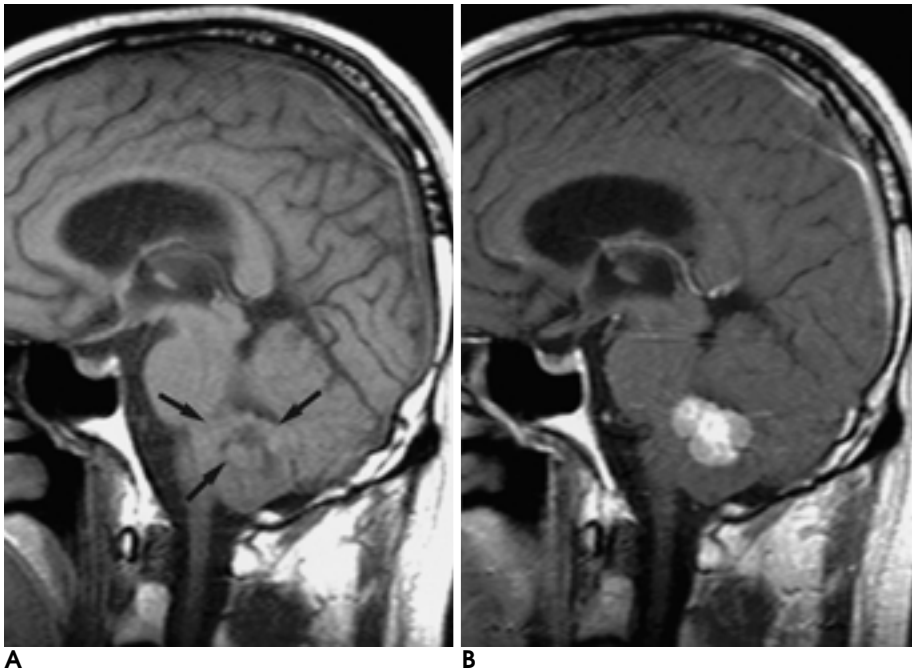


Fig. 12. Fourth ventricle metastasis in a 51-year-old man with known renal cell carcinoma.
A. At T1-weighted sagittal imaging, the mass shows mixed iso and low SI (arrows).
B. The mass shows good heterogeneous enhancement at enhanced T1-weighted sagittal imaging. He has another metastatic lesion within the right lateral ventricle (not shown).

We encountered one case involving ventricular metastasis from renal cell carcinoma (RCC) (Fig. 12); in a series of intracranial metastatic RCCs reported by Uchino et al. (14), a single fourth ventricular lesion was also present. They noted that the signal intensities of the tumors were not specific, but concluded that an intra- and/or peritumoral flow void is a fairly characteristic MRI finding of intracranial metastases of RCCs.

Conclusion

Various tumors can occur in the fourth ventricle, and this study has presented and reviewed the imaging findings of such tumors, both common and rare, which can occur in adults and adolescents. The MRI findings of the majority of these tumors were nonspecific. However, in some cases, such as ependymoma, lymphoma, epidermoid cyst, meningioma, melanoma, and cavernous hemangioma, the specific signal intensity or growth pattern observed was useful for differential diagnosis.

It is hoped that this paper will be of use to radiologists who encounter these diverse disease entities of the fourth ventricle.

References

1. Osborn AG. *Brain tumors and tumorlike masses: classification and diffrential diagnosis.* In Patterson AS, ed. *Diagnostic neuroradiology.* 1st ed. St. Louis: Mosby-Year Book, 1994:401-528
2. Taveras JM. *Intracranial neoplasms.* In Mitchell C, ed. *Neuroradiology.* 3rd ed. Baltimore: Williams & Wilkins, 1996:571-722(683)
3. Osborn AG. *Astrocytomas and other glial neoplasms.* In Patterson AS, ed. *Diagnostic neuroradiology.* 1st ed. St. Louis: Mosby-Year Book, 1994:529-578
4. Tortori-Donati P, Fondelli MP, Cama A, Garre ML, Rossi A, Andreussi L. Ependymomas of the posterior cranial fossa: CT and MRI findings. *Neuroradiology* 1995;37:238-243
5. Chiechi MV, Smirniotopoulos JG, Jones RV. Intracranial subependymomas: CT and MR imaging features in 24 cases. *AJR Am J Roentgenol* 1995;165:1245-1250
6. Sartor K. *Tumors and related conditions.* MR imaging of the skull and brain: a correlative text-atlas. 1st ed. Berlin Heidelberg: Springer-Verlag, 1992:249-494
7. Osborn AG. *Meningiomas and other nonglial neoplasms.* In Patterson AS, ed. *Diagnostic neuroradiology.* 1st ed. St. Louis: Mosby-Year Book, 1994:579-625
8. Osborn AG. *Miscellaneous tumors, cysts and metastases.* In Patterson AS, ed. *Diagnostic neuroradiology.* 1st ed. St. Louis: Mosby-Year Book, 1994:626-670
9. Lunardi P, Missori P, Gagliardi FM, Fortuna A. Epidermoid tumors of the 4th ventricle: report of seven cases. *Neurosurgery* 1990; 27:532-534
10. Ceylan S, Ilbay K, Kuzeyli K, Kalelioglu M, Akturk F, Ozoran Y. Intraventricular meningioma of the fourth ventricle. *Clin Neurol Neurosurg* 1992;94:181-184
11. Arbelaez A, Castillo M, Armao DM. Imaging features of intraventricular melanoma. *AJNR Am J Neuroradiol* 1999;20:691-693
12. Reyns N, Assaker R, Louis E, Lejeune JP. Intraventricular cavernomas: three cases and review of the literature. *Neurosurgery* 1999;44:648-654
13. Chou MS, Tsai TC, Lin MB, Liu GC, Howng SL. Metastasis involving the leptomeninges and ventricles of the brain—CT evaluation *Gaoxiong Yi Xue Ke Xue Za Zhi* 1994;10:256-262
14. Uchino A, Hasuo K, Mizushima A, et al. Intracranial metastasis of renal cell carcinoma: MR imaging. *Radiat Med* 1996;14:71-76

대한방사선의학회지 2003;49:155 - 164

성인에서의 다양한 제4 뇌실 종양: 자기공명영상 소견[1]

[1]서울대학교 의과대학 방사선과학교실

김승자 · 장기현 · 김건하 · 최자영 · 권배주 · 한문희

일차성 제4 뇌실 종양은 소아에서는 흔하지만 성인에서는 드물다. 접하게 되는 질병명의 경우에도 환자의 나이에 따라 달라진다. 저자들은 16세이상의 환자로 자기공명영상을 시행하였고 자기공명영상에서 제4 뇌실에 병변이 있었던 증례들을 수집하였다. 대부분의 환자는 절제술을 시행받았으며 병리학적으로 확진되었다. 저자들이 경험한 다양한 제4뇌실 종양들은 상의세포종 8예, 상의하세포종 1예, 맥락총 유두종 3예, 성상세포종 3예, 수모세포종 1예, 림프종 2예, 표피양낭 2예, 수막종 1예, 흑색종 1예, 해면혈관종 1예, 그리고 전이암 1예 등이었다.

저자들은 주로 제4 뇌실에 병변이 있었던 다양한 종양을 소개하고, 임상 및 방사선학적 소견을 기술할 것이며, 대부분 비특이적이었다. 그러나, 몇몇 증례에서는 특이적인 신호강도나 종양의 성장양상이 감별진단에 도움이 되었다.