MRI Findings of Intracranial Hemangioblastoma

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Purpose: Complete resection of the tumor nodule (mural nodule or solid portion of the tumor) is the essential goal of surgical treatment for hemangioblastoma. The purpose of this study was to classify the morphologic types of intracranial hemangioblastoma on MRI and to compare the location and contour of tumor nodule on MRI with those on angiography.

Materials and Methods: The MRI findings of 34 lesions (38 lesions if 4 spinal cord lesions were included) in 26 patients (17 males and 9 females, range of age, 18-67 years, mean, 39 years) with surgically and histopathologically proved intracranial hemangioblastomas were reviewed. Seventeen patients underwent CT scanning in a short interval. Contrast-enhanced T1-weighted imaging patterns of hemangioblastoma were classified according to Ho's morphologic types. The location and contour of tumor nodule were compared between MRI and angiography in 15 patients (24 lesions).

Results: By location, cerebellar hemisphere was predominated (55%), followed by cerebellar vermis (26%), supratentorial region (5%), and medulla oblongata (3%). Spinal cord lesions (11%) were seen in 3 patients of 5 von Hippel-Lindau diseases. The frequency of morphologic types was as follows: Type 1 (purely cystic), 3%, Type 2 (mural nodule), 50%, Type 3 (cyst with wall enhancement), 3%, Type 4 (cystic nodule), 15%, Type 5 (solid with internal cyst), 9%, and Type 6 (solid), 20%. All tumor nodules (33 lesions) enhanced intensely with intravenous contrast material on MRI, of which 24 lesions (in 15 patients) revealed hypervascular masses fed by pial arteries on angiography. They were superficial and abutted pia mater partially or in large portion on both MRI and angiography.

Conclusion: Over 70% of intracranial hemangioblastomas had a surrounding cyst, and superficial, pial-based location and number of the tumor nodules on MRI was correlated well with those on angiography. MRI is the examination of choice for preoperative evaluation of intracranial hemangioblastoma.

Index Words: Brain neoplasms, MR Angioma, central nervous system Cerebral blood vessels, angiography

INTRODUCTION

Hemangioblastomas are benign neoplasms of endothelial origin and account for 1-2.5% of all intracranial neoplasms. Although also found in the spinal cord (3-13%), medulla (2-3%), and cerebrum (1.5%), hemangioblastomas most commonly occur in the cerebellum (83-86%), where they comprise 7-12% of primary posterior fossa tumors. These tumors may occur sporadically or as part of the von Hippel-Lindau disease (VHLD), an inherited autosomal disorder characterized additionally by retinal angiomatis, renal cysts or carcinoma, pheochromocytoma, and pancreatic cysts or carcinoma. Cerebellar hemangioblastomas are common in middle aged adults, with the mean age in
most series between 30 and 40 years. Patients with VHLD develop cerebellar hemangioblastomas at a younger age (1-2). Approximately 70% of cerebellar hemangioblastomas are cystic, compared with only 20% of cerebral or bulbar lesion. The diagnosis of central nervous system (CNS) hemangioblastomas as well as the surgical treatment requires the accurate radiologic visualization of both cystic and solid components of the tumor, as complete resection of the tumor nodule is the essential goal of surgery (1-5).

The purpose of this study was to classify the morphologic types of intracranial hemangioblastoma on MRI and to compare, whenever possible, the location and contour of tumor nodules on MRI with those on angiography.

MATERIALS and METHODS

The MRI findings of 34 lesions in 26 patients (17 males and 9 females) with surgically proved intracranial hemangioblastomas were reviewed. Two of the 26 patients had a history of previous operation for hemangioblastoma. The patients' ages ranged from 18 to 67 years (mean, 39 years). All patients were examined on a Toshiba 0.5T MRT-50A scanner. Imaging variables in all patients included T1-weighted images (400-500/15-20/2, TR/TE/exitations) and T2-weighted images (2500-3000/30, 80) in a combination of at least two orthogonal planes. In each patient, after an intravenous injection of 0.1 mmol/kg of gadopentate dimeglumine (Gd-DTPA) (Magnevist, Shering, Germany), axial, coronal and/or sagittal T1-weighted images were obtained. Seventeen patients underwent intravenous contrast-enhanced CT scanning in a short interval. Angiograms were available for comparison in 15 patients. Radiologic patterns of the hemangioblastoma on the contrast-enhanced T1-weighted images were classified according to Ho’s morphologic types (7). The location and contour of tumor nodule (mural nodule or solid portion of the tumor) on contrast-enhanced T1-weighted images were compared with those on angiography.

Table 1. Location of CNS Hemangioblastomas

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<th>Location</th>
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<td>Supratentorial</td>
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<td>Medulla oblongata</td>
<td>1 (3%)</td>
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<td>Spinal cord</td>
<td>4 (11%)</td>
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Table 2. MRI Patterns of Intracranial Hemangioblastoma

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<th>Type</th>
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<tbody>
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<td>Type 1 (pure cyst)</td>
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<tr>
<td>Type 2 (mural nodule)</td>
<td>17 (50%)</td>
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<tr>
<td>Type 3 (cyst with wall enhancement)</td>
<td>1 (3%)</td>
</tr>
<tr>
<td>Type 4 (cystic nodule)</td>
<td>5 (15%)</td>
</tr>
<tr>
<td>Type 5 (solid with internal cyst)</td>
<td>3 (9%)</td>
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<tr>
<td>Type 6 (solid)</td>
<td>7 (20%)</td>
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<tr>
<td>Total</td>
<td>34 (100%)</td>
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</table>

Fig. 1. Vermian hemangioblastoma, typical solid nodule with surrounding cyst (type 2). Gd-DTPA enhanced axial T1-weighted images (a) reveal a small, densely enhancing “mural nodule” with an adjacent nonenhancing surrounding cyst. The lateral view of vertebral angiogram (b) demonstrates the vascular tumor nodule in the vermis in a avascular region that corresponds to the surrounding cyst (arrows).
RESULTS

The location of 38 lesions of CNS hemangioblastomas is presented in Table 1. Twenty-one lesions were located in the cerebellar hemisphere (55%), 10 lesions in the vermis (26%), 1 lesion in the medulla (3%), 2 lesions in the supratentorial (5%), and 4 lesions in the spinal cord (11%). The hemangioblastomas were multiple in 7 patients, and 5 of them had VHLD. Spinal hemangioblastomas were found in 3 of these 5 VHLD and 2 of them had a documented familial history of

Fig. 2. Cerebellar hemangioblastoma, purely solid (type 6). Axial T2-weighted image (a) reveals multiple serpentine signal voids within and at the periphery of the mass involving left cerebellar hemisphere. Gd-DTPA enhanced sagittal T1-weighted image (b) shows a large, densely enhancing nodule abutting pia mater. The lateral view of vertebral angiogram (c) demonstrates a hypervascular tumor supplied by both AICA and PICA.

Fig. 3. Cerebellar hemangioblastoma, cystic nodule (type 4). Gd-DTPA enhanced axial (a) and coronal (b) T1-weighted images reveal a large cystic nodule with an adjacent nonenhancing surrounding cyst. The A-P view of vertebral angiogram (c) demonstrates the vascular tumor with central avascular region and a draining vein.
VHLD. The tumors ranged from 0.5 to 7.8 cm in diameter.

The contrast-enhanced MRI findings of 34 intracranial lesions in 26 patients are summarized in Table 2. The most common MRI pattern of hemangioblastoma was solid "mural nodule" with an adjacent nonenhancing surrounding cyst (type 2, 50%) (Fig. 1), followed by purely solid (type 6, 20%) (Fig. 2), a cystic mural nodule (type 4, 15%) (Fig. 3), a solid mass with internal cyst (type 5, 9%), completely cystic (type 1, 3%), and a cyst with wall enhancement (type 3, 3%) (Fig. 5). Overall 71% of hemangioblastomas were predominantly cystic and the other 29% were solid. All 33 solid nodules were superficial and abutted pia mater. Serpentine signal voids within and/or at the periphery of the mass were observed in 24 lesions (70%) (Fig. 2). Cystic portion of the tumor was isointense or slightly hyperintense to CSF on all sequences. The tumor nodules were slightly hyperintense to gray matter on noncontrast T1-weighted images and long TR images and they enhanced intensely with Gd-DTPA.

Angiography showed one or more hypervascular nodules in 15 patients (24 tumor nodules), corresponding to their number and location on the contrast-enhanced MRI (Fig. 4). In 17 of 24 nodules with type 2, 3, and 4 shown on MRI, angiography defined the cyst as an avascular mass (Fig. 1). The tumor nodules stained intensely, either with a homogenous or mottled appearance, and were associated with one or more enlarged feeding (pial) arteries (superior cerebellar artery, anterior inferior cerebellar artery, posterior inferior cerebellar artery, middle cerebral artery, and occipital artery of external carotid artery) (Fig. 1, 2, 3, 4). Six out of 24 hypervascular nodules had 2 or 3 feeding arteries (Fig. 2) and early draining vein was seen in 10 lesions (Fig. 3).

**DISCUSSION**

Hemangioblastomas originate from the pia, grow slowly, and then subsequently develop either into a well-circumscribed large cystic tumor or a well-circumscribed solid tumor (1, 4-6). Morphologically, hemangioblastomas are pink to yellow, and abut the pial surface of the cerebellum, where the solid nidus (mural nodule or solid portion of the tumor) is found (1-13). Dural involvement is present in up to 20% of posterior fossa hemangioblastomas (11). The "cyst" around the mural nodule is a collection of fluid arising by diffusion from the vascular component of the mural nodule and is not truly part of the neoplasm (1-2, 11). The cyst wall, if present, is composed of compressed adjacent brain parenchyma or reactive neuroglial cells. The cysts may also occur within a mural nodule or solid tumor and in such case the cysts are part of the tumor representing dilated vascular spaces or necrosis within the neoplastic tissue of the hemangioblastoma (1, 10-13) (Fig. 3).

Effective surgical treatment of CNS hemangioblastoma requires complete excision of the tumor nodule. A safe approach and resection are best accomplished when the exact location of the nodule and its relationship to the tumor cyst and adjacent structures are
known(3, 5, 14–15). MRI is superior to CT in delineating the exact extent of the hemangioblastomas, for example contact with the arachnoid surface. Connection of tumors to the surarachnoid space arises from the tumor’s origin in the pial mesenchyma(8–9).

Characteristic CT findings of a posterior fossa hemangioblastoma include a cystic lesion with a small mural nodule, that intensely enhances following intravenous contrast medium administration. Alternatively, the lesion may be solid and present as a densely enhancing nodule without cyst formation(16–18). The primary limitations of CT include decreased posterior fossa sensitivity, compared with MRI, due to artifacts from the base of the skull.

The MRI characteristics of hemangioblastoma are: (1) the cystic nature of the mass, (2) a peripheral, pial-based mural nodule of solid tissue that enhances markedly with intravenous contrast, and (3) large vessels within and/or at the periphery of the mass(serpentine signal voids) suggestive of dilated feeding or draining vessels that accompany these hypervascular tumor. These are virtually pathognomonic for hemangioblastoma when found in conjunction with each other(7, 10, 19–23). The cysts are sharply margined and smoothly bordered and isointense or slightly hyperintense to CSF on all sequences. The mural nodule or solid portion of the tumor is only slightly hyperintense to gray matter on noncontrast T1-weighted images and long TR images. Entirely solid hemangioblastomas occur in 30–40% of cases and are most common morphologic type if in the supratentorial compartment(7, 10, 15, 24–25). The 20% of solid tumors in our series is less in frequency than that reported in the literature. Seven cases of supratentorial intraventricular hemangioblastomas have been described in the literature and interestingly, all of these have been solid tumors(25). Supratentorial hemangioblastomas are extremely rare. Using data from previous reports there are 82 cases of supratentorial hemangioblastomas in the literature(25). In our study 2 of 38(5%) CNS hemangioblastomas occurred in the cerebrum and both cases(100%) were cystic, which was higher in frequency than that of previous report(20%) although the number of subjects was smaller in this study. On MRI 60–69% of hemangioblastomas have associated internal and/or peripheral serpentine signal voids(7, 10, 26), which was similar in frequency to our study(70%).

Ho et al. described 6 radiologic types of hemangioblastomas(including the frequency in each type) as visualized on MRI and CT(7)(Fig. 6): (1) purely cystic (type 1, 8%), (2) solid “mural nodule” with an adjacent nonenhancing surrounding cyst(type 2, 35%), (3) a mural nodule associated with an enhancing cyst wall(type 3, 6%), (4) cystic nodule(type 4, 6%), (5) a solid mass
with internal cysts (type 5, 12%), and (6) purely solid (type 6, 33%). Overall, roughly 55% of hemangioblastomas had surrounding cysts and the other 45% were predominantly solid and this radiographic distribution correlated well with the pathologic spectra of 60% cystic and 40% solid described by Rubinstein (1). Our study showed slightly higher incidence (71%) of intracranial hemangioblastomas with a surrounding cyst (type 1 3%, type 2 50%, type 3 3%, type 4 15%). The most common MRI pattern was type (50%) as was in Ho's report (35%). All tumor nodules (type 2, 3, 4, 5, 6) were superficial and abutted pia mater. The cyst wall usually does not enhance. However, if the cyst is lined by neoplasm, the wall will enhance (7, 10, 26-27). Only one case showed a cyst with wall enhancement in our study (Fig. 5). Tumor cells were demonstrated histopathologically in this enhancing wall.

As the differential diagnosis of hemangioblastoma on MRI, cystic astrocytomas and meningiomas are essential. The mural nodule abuts a pial surface in the cystic hemangioblastoma. In contrast, the mural nodule of cystic astrocytoma is found within the cerebellar parenchyma at a distance from a pial surface. Solid hemangioblastoma can be differentiated from meningioma by the shape of the "flow voids" around the tumor. The former appears as the spotty or serpentine shape and the latter as the smooth peritumoral rim (1, 8). Guhl et al. (8) reported another way to differentiate between these tumors by the difference of the T2 relaxation time; the T2 values of angiomatosus tumor were greater than those of meningioma. Supratentorial hemangioblastomas presenting as a cyst with mural nodule, abutting on the falx but not adherent to it, provides the differential diagnosis with cystic astrocytoma and angioblastic meningioma. The nature of enhancement excludes the astrocytoma. Hemangioblastomas are round and have no flat base on the arachnoid, unlike an angioblastic meningioma. It has been accepted that dural attachment does not preclude the diagnosis of hemangioblastoma. Tentorial branches feeding a supratentorial hemangioblastoma have been described (4, 16, 28). In our study there was no difficulty in the diagnosis of hemangioblastomas on MRI except a calcified supratentorial lesion.

Angiography provides detection and exact localization and the vascularization of the hemangioblastomas in the distribution of the vertebrobasilar system, but rarely from branches of the carotid arteries (meningeval tentorial and ascending pharyngeal arteries). Hemangioblastomas are vascular tumors that have a dense tumor nodule or a heterogenous network of tangled vessels fed by a dilated artery and sometimes draining veins. The latter case occasionally erroneously suggests an arteriovenous malformation. If the tumor sits within a cyst, it appears as a vascular nodule within an avascular region, secondary to displacement of the surrounding vessels by the avascular cyst. Additionally, angiography is very useful in the demonstration of multiple tumors and above all of the small tumors, which sometimes are not diagnosed by CT scanning and noncontrast MRI (7, 10, 14, 26). With noncontrast MRI the tumor nidus may blend with surrounding edema and may not be visualized. Limitations of noncontrast MRI also preclude detection of small hemangioblastomas that are not accompanied by mass effect, edema, or cyst (7, 10, 19-21, 26). In our study the number of hypervascular nodules or masses detected on angiography was identical to that of the enhancing nodules on MRI. All tumor nodules of hemangioblastoma abutted pial surface on MRI and were fed by the pial arteries angiographically.

In conclusion, over 70% of the intracranial hemangioblastomas had surrounding cysts, and superficial pial-based location and number of the tumor nodules on contrast-enhanced MRI were correlated well with those on angiography. With above mentioned MRI findings, intracranial hemangioblastomas can be easily diagnosed by contrast-enhanced MRI in most cases and angiography will be necessary to identify the vascular supply of the lesion before surgery.

REFERENCES

혈관아세포종의 자기공명영상 소견

목 적: 혈관아세포종의 치료목적은 종앙결절(벽결절이나 종괴의 고형부분)을 완전히 제거하는 것이다. 뇌 혈관아세포종의 조영증강후의 T1강조영상 소견을 형태학적으로 분류하고, 종앙결절의 위치와 양상을 혈관촬영 소견과 비교하고자 하였다.

대상 및 방법: 수술로 확진된 뇌 혈관아세포종 34병소(26명의 환자, 남자 17명, 여자 9명, 연령분포 18-67세, 평균연령 39세)의 조영증강후의 T1강조영상 소견을 Ho 등의 분류에 따라 형태학적으로 분류하였고, 혈관촬영을 시행한 15명의 환자(24병소)에서 종앙결절의 위치와 양상을 조영증강후 T1강조영상 소견과 비교하였다.

결 과: 발생부위별로는 소뇌반구가 가장 많았고(55%), 다음으로 소뇌충수(26%), 전막상부(5%), 연수(3%)의 순이었으며, 척수병변(11%)은 von Hippel-Lindau 병 환자 중 3명에서 나타났다. 조영증강후의 T1강조영상 소견의 형태학적 분류는 제1형(순수한 낭종형)이 3%, 제2형(벽결절을 가진 낭종형)이 50%, 제3형(조영증강되는 벽을 가진 낭종형)이 3%, 제4형(낭성의 벽결절을 가진 낭종형)이 15%, 제5형(내부 낭종을 가진 고형형)이 9%, 그리고 제6형(고형성)이 20%이었다. 종앙결절 모두가(33병소)가 조영제 주입 후에 강한 조영증강을 보였는데 이중 혈관촬영을 시행한 24병소는 뚜렷한 과혈관성 종괴를 나타내었고 연막동맥들로부터 공급받고 있었다. 또한 이들 24병소는 표재성으로써 일부 또는 대부분을 연막에 기저 두고 있는 양상을 조영증강후의 T1강조영상과 혈관촬영상에서 일치하였다.

결 론: 뇌 혈관아세포종의 70%이상이 낭종형이었으며, 조영증강후 T1강조영상과 혈관촬영상 종앙결절의 위치와 양상이 일치하였다. 고로 뇌 혈관아세포종의 숙전 진단에는 조영증강영상이 필수적 방법이라고 할 수 있다.
1996년도 의사전문의고시 문제 출제경향 안내

1. 전문의 시험 분야별 출제비율

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2. 핵의학 분야 수련 및 출제
핵의학 분야의 수련은 현행대로 2개월 이상의무적으로 시행해야 하며 전문의 시험에도 핵의학을 현행 비율대로 계속 출제 할 것임.

3. 동위원소 취급 특수면허 취득을 위한 교육이나 동면허취득으로 상기 2항의 수련 의무를 대신하지 못함.

4. 방사선 관계 법규를 출제율 수에서 1% 전후 출제 할 것임.

5. 상기 출제 비율은 당해년도 문제선택위원의 성향 또는 문제운행의 문제성향 등에 따라서 증감이 될 수 있음.