

MR Findings of Wernicke Encephalopathy*

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— Abstract —

Seven patients (33 to 58 years old) with clinical diagnoses of Wernicke encephalopathy were examined with MR on either a 2.0T (5 cases) or a 0.5T scanner (2 cases) using spin-echo pulse sequences. In 2 patients, follow-up MR studies were performed 1 and 5 weeks after thiamine (vitamine B1) treatment.

Five patients (4 chronic alcoholics and 1 with hyperemesis gravidarum) showed atrophy of both mamillary bodies, along with patchy lesions around the third ventricle, medial thalami, tectum of the midbrain, and periaqueductal gray matter. Another patient with hyperemesis of gravidarum demonstrated only slightly atrophic mamillary bodies, and the last patient with severe vomiting after gastrojejunostomy showed only diencephalic/mesencephalic lesions with apparently normal mamillary bodies. A follow-up MR showed a decrease in previously-noted diencephalic-/mesencephalic lesions but no change in the size of the mamillary bodies. Diencephalic/mesencephalic lesions were well seen as a high-signal intensity on proton- and T2-weighted axial images, while atrophy of the mamillary bodies was seen best on T1-weighted sagittal images.

MR imaging is very useful in demonstrating the characteristic lesions of Wernicke encephalopathy and in evaluating the result of treatment on follow-up study.

Index Words: Brain, abnormality

Brain, MR studies 10.1214

Wernicke encephalopathy

Wernicke encephalopathy is a neurologic complication of chronic alcoholism and malnutrition caused by a nutritional deficiency of thiamine (vitamin B1). It is characterized by clinical triad of mental symptoms, paralysis of the extra-ocular muscles, and ataxia. It usually presents acutely and is often fatal condition, but, which is treatable and preventable with adequate thiamine supplementation (1).

The main pathologic changes consist of macro- or microscopic mamillary body abnormalities (shrinkage or petechial hemorrhage), and edema, gliosis, degeneration or demyelination associated

with endothelial proliferation or hemorrhage around the third ventricle, periaqueductal gray matter and medial dorsal nuclei of the thalami (2-6).

There have been only a few neuroimaging studies of Wernicke encephalopathy reported in the literature (7-13). The purpose of this article is to describe the magnetic resonance (MR) imaging findings of seven cases with Wernicke encephalopathy.

Materials and Methods

Seven patients with clinical diagnoses of Wernicke encephalopathy (five men and two women, 33-58

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years old), were examined with MR during 13 month-period (February 1990 to February 1991) at Seoul National University Hospital and Young-Deung-Po City Hospital, Seoul, Korea. Four patients were chronic alcoholics and three were nonalcoholics (two with hyperemesis gravidarum and one with prolonged vomiting after gastrojejunostomy due to duodenal ulcer). All seven patients had classical triad of clinical symptoms (mental disturbances, ophthalmoplegia and ataxia) which improved after intravenous or oral thiamine administration.

MR studies were performed using either a 2.0 Tesla scanner (Goldstar, Spectro-2000) (5 cases) or a 0.5 Tesla scanner (Goldstar, Supertec-5000) (2 cases) using spin-echo pulse sequences (T1-weighted images: TR = 500-600 msec, TE = 25-30 msec, proton density- and T2-weighted images: TR = 2300-2500 msec, TE = 30 and 70-100 msec) before the initiation of treatment. Acquisition matrix was 256 × 200-256 and field of view was 22-23cm. The slice thickness and gap were 5-7mm and 2-3mm, respectively. Number of excitation (average) was 2-5 for T1-weighted images and one for T2-weighted images. Routine imaging planes were sagittal for T1-weighted images and axial for T2-weighted images. coronal images were additionally obtained in three patients. Two patients had follow-up studies 1 week (case 4), and 1 and 5 weeks (case 3) after the initial study.

MR findings were retrospectively reviewed with special attention to presence or absence of mamillary

body atrophy, and abnormal signal intensities around the third ventricle, periaqueductal gray matter and thalami. Associated findings such as cerebral or cerebellar atrophy, corpus callosal atrophy, etc. were also evaluated. The mamillary bodies were designated to be "atrophic", when they were invisible on both sagittal T1-weighted and axial T2-weighted images.

Results

Clinical presentations and MR findings are summarized in Table 1.

Mamillary bodies were invisible on both T1-weighted sagittal and T2-weighted axial images, suggesting atrophy in 5 cases. In one of the two remaining cases, the mamillary bodies appeared to be slightly smaller than usual. The remainder showed normal size of mamillary bodies (Fig. 1). Bilateral symmetrical patchy hyperintense lesions were demonstrated around the third ventricle, periaqueductal gray matter and/or medial portions of the thalami on T2-weighted images in 6 cases (Figs. 1,2,3,4). On T1-weighted sagittal images, the lesions were difficult to be identified.

Five patients including all 4 alcoholics presented both mamillary body atrophy and hyperintense lesions in the mesencephalic/diencephalic areas (case 2,3,4,6,7). One patient with hyperemesis gravidarum showed mamillary body atrophy but no hyperintense lesions in the mesencephalic/diencephalic areas

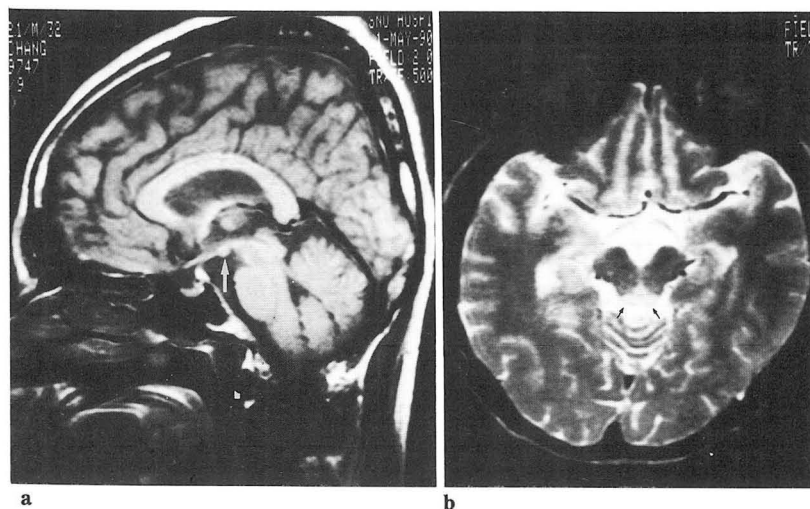


Fig. 1. Case 1

a. Mamillary body appears normal in size on T1-weighted (SE500/30) mid-sagittal image (arrow).

b. T2-weighted (SE2500/80) axial image shows hyperintense lesions in periaqueductal gray matter and tectum of the midbrain (arrows).

Table 1. Clinical Presentations and MR Findings.

Age/ Sex	Clinical presentations				MR findings			Clinical course
	Chronic alcoholics	Mental state	External ocular movement	Gait	Mamillary body	Mesen-/diencephalic lesions	Associated findings	
Case 1 35/M	- (s/p gastro- jejunostomy)	confusion short term memory loss	bilateral horizontal gaze palsy with vertical nystagmus	truncal ataxia	normal	periaqueductal gray matter around third ventricle bilateral medial thalami tectum of midbrain		improved all sxs. except memory loss
Case 2 48/M	+	drowsy	complete external ophthalmoplegia	inability to stand alone	atrophic	periaqueductal gray matter around third ventricle bilateral medial thalami	cerebellar atrophy	full recovery after 12 days
Case 3 58/M	+	confusion	bilateral medial gaze palsy with exotropia	inability to stand alone	atrophic	periaqueductal gray matter around third ventricle (gradually decreased on F/U MR 1- and 5 weeks later)	cerebral atrophy corpus callosal atrophy	improved all symptoms
Case 4 36/F	- (hyperemesis gravidarum)	drowsy short term memory loss	gaze evoked nystagmus	inability to stand alone	atrophic	periaqueductal gray matter around third ventricle bilateral medial thalami tectum of midbrain (decreased on F/U MR 1 week later)	cerebral atrophy subcortical hyperintense lesions	improved all symptoms
Case 5 33/F	- (hyperemesis gravidarum)	confabulation short term memory loss	gaze evoked nystagmus	inability to walk alone	slightly atrophic	normal	cerebral atrophy subcortical hyperintense lesions	improved all sxs. except memory loss
Case 6 58/M	+	drowsy	mild bilateral horizontal gaze palsy	inability to walk alone	atrophic	periaqueductal gray matter around third ventricle	cerebral atrophy cerebellar atrophy corpus callosal atrophy	full recovery after 10 days
Case 7 45/M	+	drowsy	bilateral medial gaze palsy with nystagmus	inability to stand alone	atrophic	periaqueductal gray matter around third ventricle		full recovery after 10 days

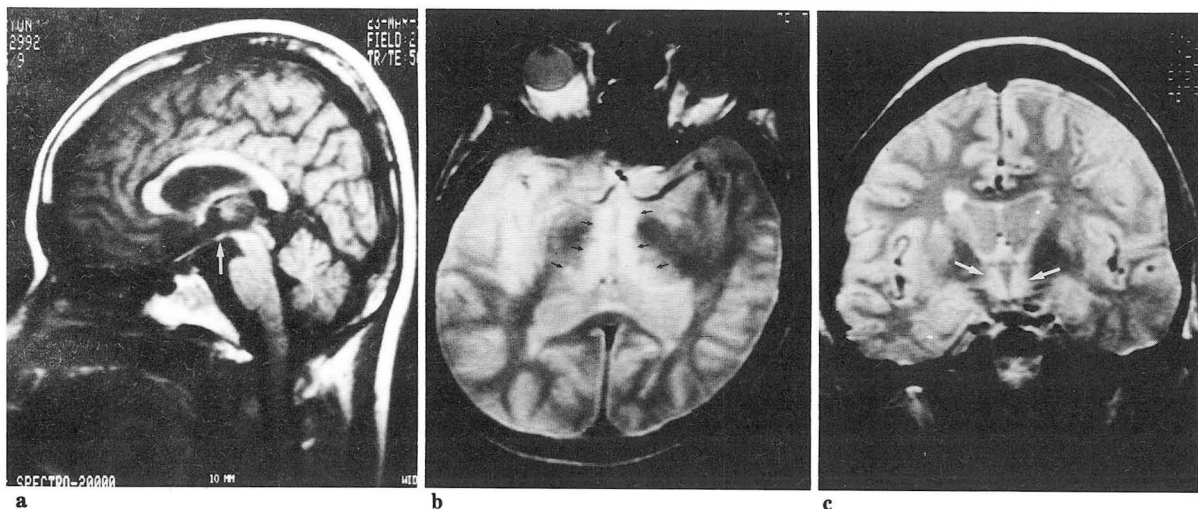


Fig. 2. Case 2

a. T1-weighted (SE500/30) mid-sagittal image shows atrophy of the mamillary bodies (arrow).
b & c. Proton-density (SE2500/30) axial (b) and coronal (c) images show hyperintense lesions around the third ventricle and in the medial dorsal thalamic nuclei (arrows).

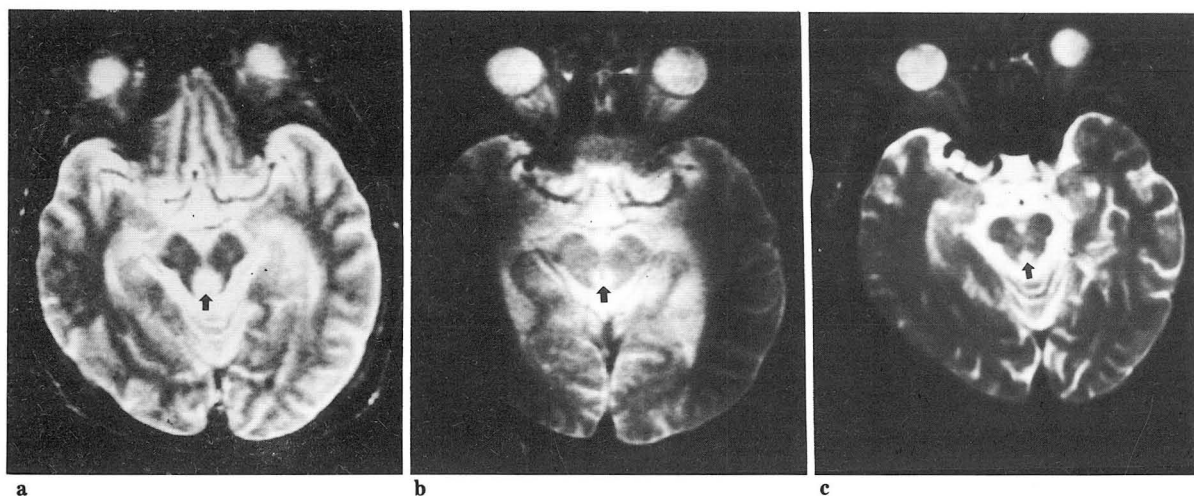


Fig. 3. Case 3

a, b & c. T2-weighted (SE2500/80) axial image (a) shows large hyperintense lesion around the aqueduct (arrow), which hyperintense lesions around the third

(case 5), while another patient with prolonged vomiting after gastric surgery demonstrated the hyperintense mesencephalic lesions but normal mamillary bodies (case1).

Other associated findings included cerebral cortical atrophy (4 cases), cerebellar atrophy (2 cases), corpus callosal atrophy (2 cases) and subcortical patchy hyperintense lesions (2 cases).

Follow-up MR studies performed after clinical symptoms improved with vitamin B1 administra-

tion in two patients showed decrease of the hyperintense lesions on T2-weighted images in both size and signal intensity but no change of mamillary body atrophy (Figs. 3,4).

Discussion

Since its original description in 1881, Wernicke encephalopathy has become recognized as a disorder of thiamine deficiency closely associated with chronic

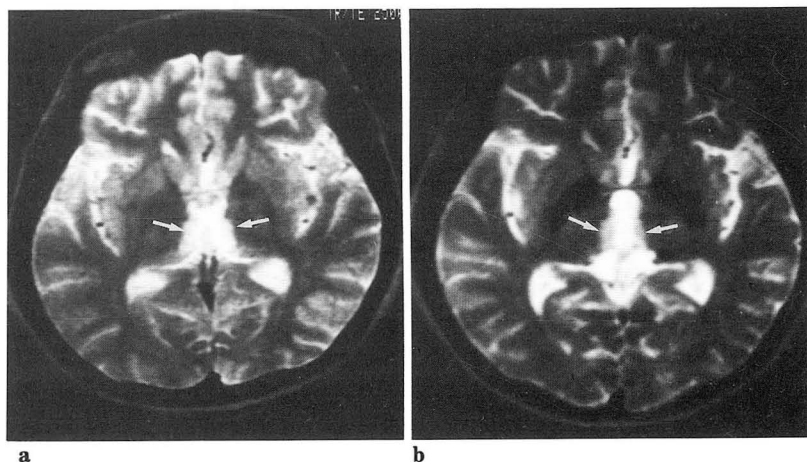


Fig. 4. Cases 7

a & b. T2-weighted (SE2500/80) axial image (a) shows symmetrical hyperintense lesions around third ventricle (arrows). Follow-up SE2500/80 axial MR image 1 week later (b) demonstrates decrease of the lesions (arrows).

alcoholism. It may also complicate other various conditions including hyperemesis gravidarum, anorexia nervosa, prolonged intravenous feedings, hemodialysis, neoplasm, gastric plication surgery, etc (14-22).

A precise pathogenesis is still not completely understood, but possible mechanisms include metabolic disturbances (related to reduced levels of thiamine requiring enzymes, transketolase and pyruvate decarboxylase, and decreased ATP synthesis), neurophysiological disturbances (due to abnormality of neuronal membrane function and serotonergic system), and genetic predilection (it develops in only a small minority of alcoholics and other chronically malnourished persons) (2,3,6,14). The lesions are typically located in the periventricular regions of the thalamus and hypothalamus, periaqueductal regions of the midbrain, and fourth ventricle floor because these regions are more vulnerable to thiamine-deficient condition (4). The most constant change is noted in the mamillary body. Although gross atrophic changes of the mamillary bodies are encountered in about 60%, the mamillary bodies almost always show microscopic abnormalities such as cellular edema or petechial hemorrhage. The pathologic changes around the third and fourth ventricles, periaqueductal gray matter, and the thalami consist of cellular edema and vascular endothelial proliferation in acute phase, and degeneration, demyelination and gliosis in chronic phase (4,5,20,21,22).

The diagnosis of Wernicke encephalopathy has

been generally based on a combination of history, clinical symptoms and laboratory results. In many cases, the disease may progress very slowly; thus they are misdiagnosed during their life time (15,20,21,22). Since it is a treatable and even preventable disease, earlier diagnosis is of the utmost clinical importance to reduce the morbidity and mortality.

There have been a few case reports on computed tomographic (CT) findings of Wernicke encephalopathy, but CT is neither sensitive nor specific to display above mentioned pathologic changes (7,8,9).

Galluci et al reported MR findings of Wernicke encephalopathy in five patients in whom they observed the typical locations of the lesions, i.e. mamillary body, periaqueductus, brain stem and around the third ventricle which well correlated with the result of the autopsy studies (9). MR of our present cases also showed the lesions similar to those. Nonvisualization of mamillary bodies on MR, particularly on T1-weighted sagittal images, is thought to usually indicate the macroscopic atrophy of the mamillary bodies, even normal mamillary bodies can not be visible due to partial volume averaging. In our experience, sagittal T1-weighted images almost always demonstrated normal mamillary bodies, even with slice thickness of 7mm. In the present series, one patient showing apparently normal mamillary bodies on MR might have microscopic abnormalities as described in the autopsy studies (20,21,22). The patient with no visible abnormal MR findings in the mesencephalic/diencephalic areas (case 5) also

might have some microscopic abnormalities. Other findings such as cerebral cortical atrophy, cerebellar atrophy, corpus callosal atrophy and subcortical patchy hyperintense lesions, which were usually observed in chronic alcoholic patients, were not constant and not specific findings of Wernicke encephalopathy (23,24,25).

It is concluded that MR is highly sensitive and specific for the diagnosis of Wernicke encephalopathy, and is very useful particularly when the symptoms are obscure or the patients are comatous. Follow-up MR is also valuable for assessing the effectiveness of treatment after thiamine treatment.

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〈국문요약〉

베르니케 뇌증의 자기공명영상 소견

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베르니케 뇌증(Wernicke encephalopathy)은 만성 알코올 중독 등에서 비타민 B1 결핍에 의해 발생하는 드문 신경계 질환으로서 의식 장애, 안구 이상 운동, 보행 장애의 특징적 임상증세를 나타낸다. 저자들은 최근 13개월간 서울대학교병원 및 시립영등포병원에서 베르니케 뇌증으로 진단된 7례의 자기공명(MR) 영상 소견을 관찰 분석하였다. 선행질환으로 4례는 만성 알코올 중독이었으며 2례는 임신오조, 다른 1례는 위공장문합술 후 발생한 심한 구토증이었다. MR촬영은 2.0T(5례) 혹은 0.5T(2례) 초전도형 장치를 사용하였으며 스핀 에코 방법으로 T1 강조영상, 양자밀도 강조 영상 및 T2 강조 영상을 얻었다. 2례에서 비타민 B1 투여 후 각각 1주 후, 1주 및 5주 후에 추적 MR을 시행하였다.

만성 알코올 중독 환자 4례를 포함한 5례에서 유두체 위축 및 간뇌/중뇌 병변이 발견되었고, 임신오조 환자 1례에서는 경미한 유두체 위축만이, 위공장문합술 환자에서는 간뇌/중뇌 병변만이 관찰되었다. 유두체 위축은 T1 강조 시상영상에서 유두체가 보이지 않는 소견으로 나타났고, 간뇌/중뇌 병변은 T2 강조 영상에서 제3뇌실, 제4뇌실, 뇌수로관 주위 혹은 시상의 내측에 대칭성 고신호 강도로 나타났다. 추적검사를 시행한 2례에서 간뇌/중뇌 병변의 뚜렷한 호전(감소)을 보였으나 유두체의 변화는 없었다.

결론적으로 MR영상은 베르니케 뇌증의 조기진단 및 확진에 도움을 줄 뿐아니라, 추적검사로써 치료효과 판정에도 도움을 주는 유용한 검사 방법이라 사료된다.