MR IMAGING OF THE BRAIN IN WILSON'S DISEASE.

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ABSTRACT

Four cases of proven Wilson's disease underwent MRI studies of brain. The characteristic appearance of the putaminal lesion with high and low intensity spots centrally and high intensity "frame" peripherally was shown on T2WI.-TSE in three neurologically symptomatic patients. We also found that the gradient echo imaging showed low signal intensity not more pronouncely than the spin echo imaging. The mid brain,brain stem and cerebellar lesions were also seen in symptomatic patients. Some good correlation of the neurological examinations and MRI findings were found.

Key Words: Wilson's disease, MR.Imaging.

Wilson's disease or hepatolenticular degeneration is an uncommon autosomally recessive inherited disorder of copper metabolism characterized by abnormal deposition of copper in various tissues,most notable in brain and liver. Although it is rare,early diagnosis and prompt treatment can prevent the devastating neurologic sequelae of the untreated illness. In this report,we described the magnetic resonance (MR) findings in various pulse sequences and some clinical correlation of the patients.

MATERIALS AND METHODS

We have studied four male patients with clinically and biochemically proven of Wilson's disease. The age of the patients are between 11 and 24 years. The magnetic resonance images (MRI) of the brain were done with 1.5 Tesla magnet (Philips Gyroscan). In all patients,axial scan was done in T1WI and dual T2WI-TSE (Turbo-spin echo) (TR/TE =450/15;2000/25,120). The TSE is the fast spin echo technique in this machine. The sagittal T1WI-TSE and coronal T2WI-TSE were also obtained. The axial fast field echo (FFE) was also done in three of the four patients (TR/TE/FA =600/15/25). The FFE is the gradient echo imaging in this machine.

RESULTS

Three of the four patients have neurological symptoms and signs as shown in Table 1. The fourth patient came to the hospital with liver disease and abnormal liver function test but no abnormal neurological symptom and sign.

The MRI findings are also summarized in the table. All patients who have neurological deficits show abnormalities on MRI. The common finding is the abnormality of both basal ganglions especially the lentiform nucleus. In the center of the putamen,there is spots of hypointensity on both T1WI and T2WI interspersed with spots of high intensity on T2WI. The periphery shows a band of high intensity surrounding the putamen appeared to "frame" the putamen (Fig.1A).Only one case shows abnormal high intensity in the globus pallidus on T2WI (Fig.2). The caudate nucleus was involved in varying degree in all three patients.

In gradient echo imaging of the two symptomatic patients,the abnormal low signal intensity in the putamen appears not more pronoucedly than that as shown on T2WI-TSE (Fig.1B). In the asymptomatic patient,the GEI reveals no abnormality as well as T2WI-TSE (Fig.3).

All symptomatic patients have abnormal

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GEI = Gradient Echo Imaging

high intensity in the mid brain or lower brain stem,but no good correlated symptoms can be done. The patient who has rigidity and bradykinesia (Case #2) has abnormal high intensity in the dentate nucleus as well as the red nucleus and periaqueductal gray matter,while the patient who has lesion in mid brain (Case #1) shows no other neurological symptoms except dysarthria.

DISCUSSION

Wilson's disease was first described by Kinear Wilson in 1912. The pathological changes in the central nervous system are always associated with increased tissue copper content. The toxic copper substances interfere with enzymatic activity or damage cellular structure.

Many articles have dealt with computed tomography and MRI findings in the brain in Wilson's disease. The most common described abnormality is the lesion in basal ganglion. In the earlier studies of MRI using low field magnet,only high intensity was noted in the affected parts of the brain[1]. In the later articles,high field MRI have also shown low intensity in the lesion.[4,5,6,9] The typical findings on MRI are high intensity in the putamen,caudate nucleus and globus pallidus bilaterally.[1,3,4,5,6,7,8] In the putamen,low intensity of the lesion seen on T2WI but high intensity band in the periphery,mainly lateral,appeared to frame the the putamen have been described.[4,7] In our cases,there is also spots of high intensity interspersed with low intensity in the center of the putamen. The possible pathologic explanation is the cavitating degeneration of the neuronal tissue with deposition of iron and/or copper.[5,6]

The susceptibility effect is more pronounced

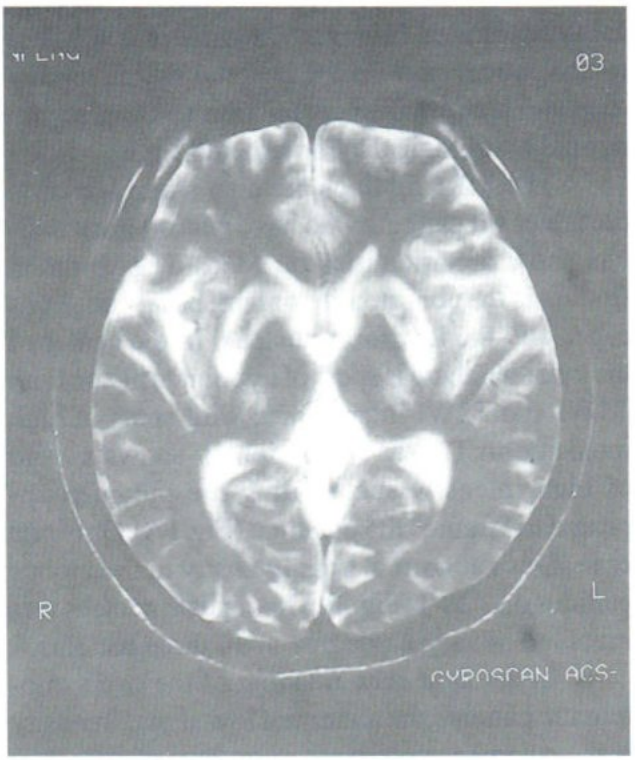


Fig.1A Axial T2WI of the patient No.1 shows characteristic hypointensity and hyperintensity lesion in the putamen and caudate nucleus. Notethe "frame" appearance of the putamen.

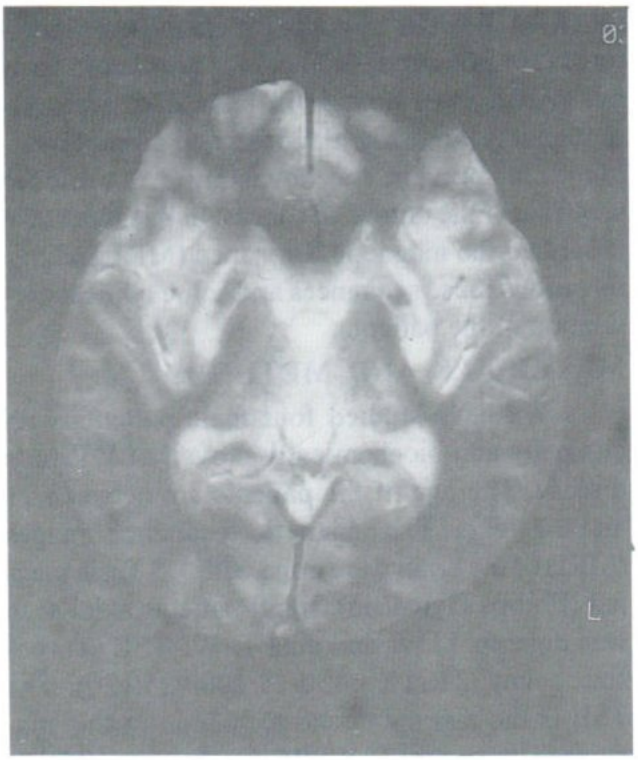


Fig. 1B Axial GEI of the same level shows the same changes of the basal ganglions.

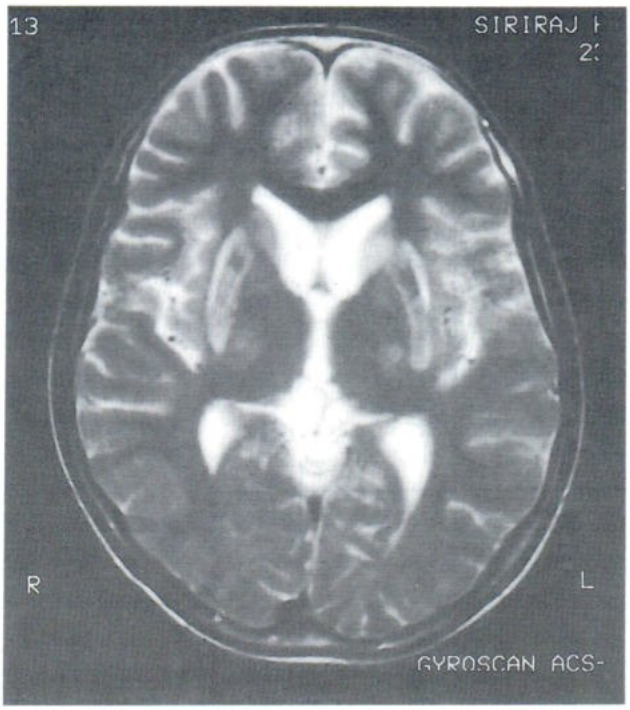
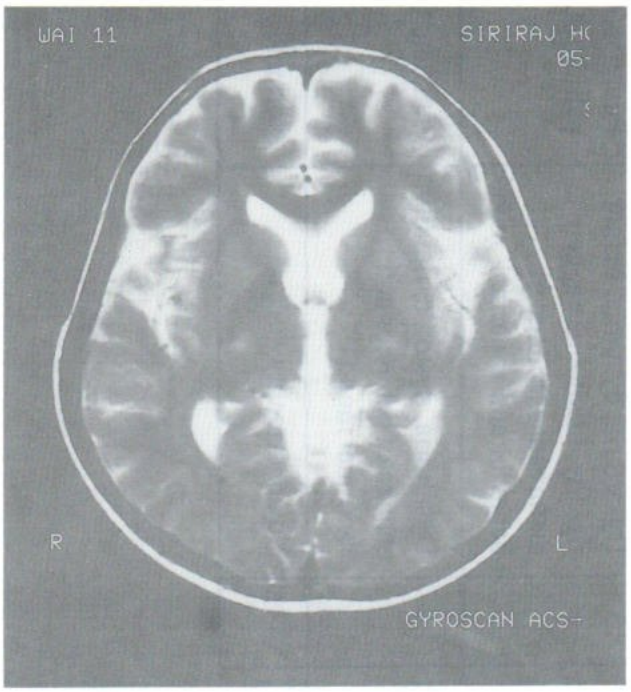


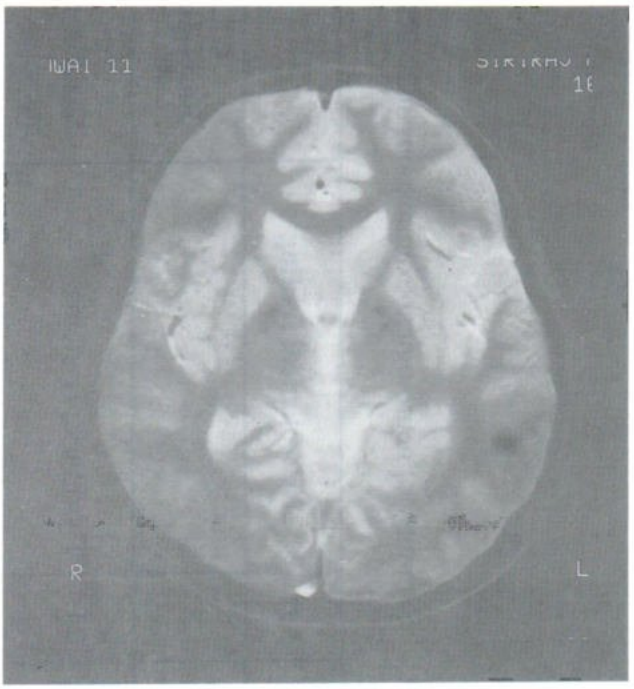
Fig. 2 Axial T2WI of the patient No.2 shows abnormal signal intensity in the basal ganglions. Note the high intensity in the globus pallidus.

in increasing magnetic field,compartmentalization of the paramagnetic substances,prolonged TE,and gradient echo pulse sequences. The blooming appearance in the paramagnetic deposition area is the suggestive evidence. In our cases,we noticed no blooming effect on gradient echo images. The explanation of low intensity may be not simply intracellular accumulation of the paramagnetics. Eventhough there are many reports about the low intensity on T2WI,no report has been done on gradient echo images to detect the deposition of paramagnetics in the basal ganglion of the Wilson's disease. We think that more cases are needed to evaluate the benefit of the gradient echo imaging in detecting very early copper or iron deposition in the brain before seen by spin-echo imaging. However,in our study it seems to be likely that,the gradient echo images will not be more sensitive in this matter.

Aisen et al[1] studied 23 patients with Wilson's disease and found good correlation between the neurological examination and the MRI findings. Many articles were also reported the same result.[3,4,7] In our study we found some



A



B

Fig.3 (A,B) : Axial T2WI and GEI of the patient No.4 shows no abnormal intensity in the basal ganglion.

Table 1 Summary of neurological examinations and MRI findings.

Patient	Age	Neurological finding	Abnormal MRI			
			Basal ganglion	**Mid brain**	**Cerebellum**	**Ventricular and subarachnoid**
1	24	Dysarthria, Dystonia	Putamen, Caudate nucleus	Red nucleus, Substantia nigra, Periqueductal gray	Normal	Mild dilatation
2	13	Dysarthria, dystonia, tremor, rigidity, bradykinesia, gait disturbance, change of behavior and abnormal mood.	Putamen, Globus pallidus, Caudate n.	Red n., Periaqueductal gray	Dentate n.	Normal
3	12	Dysarthria, dystonic, rigidity, change of behavior and abnormal mood	Putamen, Caudate n.	Normal	Cerebellar peduncle	Mild to moderate dilatation
4	11	No abnormality	Normal	Normal	Normal	Mild dilatation

correlation of the clinical and MRI findings. We also found abnormality in the mid brain, lower pons and cerebellum. The severity of the neurological examination cannot be explained by the only findings of the lesion in the brain stem, suggestive of the significant cooperative function of the supratentorial, brain stem and infratentorial neural tissue.

REFERENCES

1. Aisen AM., Martel W., Gabrielsen TO., et al (1985) Wilson's disease of the brain:MR Imaging. Radiology 157:137-141.
2. Imiya M., Ichikawa K., Matsushima H., et al (1992) MR of the base of the pons in Wilson disease. AJNR 13:1009-1012
3. Oder W., Prayer L., Grimm G., et al (1993) Wilson's disease:evidence of subgroups derived from clinical findings and brain lesion. Neurology 43:120-124.
4. Magalhaes ACA., Caramelli P., Menezes JR., et al (1994) Wilson's disease:MRI with clinical correlation. Neuroradiology 36:97-100.
5. Sener RN. (1993) Wilson's disease:MRI demonstration of cavitations in basal ganglion and thalami. Pediatr. Radiol. 23:157.
6. Brugieres P., Combes C., Ricolfi F., et al (1992) Atypical MR presentation of Wilson disease:possible consequence of paramag netic effect of copper? Neuroradiology 34:222-224.
7. Prayer L., Wimberger D., Krammer G., et al (1990) Cranial MRI in Wilson's disease. Neuroradiology 32:211-214.
8. Hitoshi S., Iwata M., Yoshikawa K. (1991) Midbrain pathology of Wilson's disease:-MRI analysis of the three cases. J. of Neurosurg., Psych. 54(7):624-6.
9. Yuh WT., Flickinger FW. (1988) Unusual MR findings in CNS Wilson disease (letter). AJR 151:834.