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MAGNETIC RESONANCE IMAGING OF EXTRAPYRAMIDAL DISEASES

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INTRODUCTION

In general, the MR brain findings in patients with extrapyramidal diseases are unspecific and consist of general or focal atrophy, iron deposition in the basal ganglia and, of regions of altered signal intensity in various extrapyramidal nuclei. Since MRI demonstrate unspecific findings, frequently observed changes of the elderly brain may cause some problems in the differentiation of extrapyramidal specific lesions and other findings unrelated to the movement disorder.

The first part of this contribution is dealing with general neuroimaging patterns in the elderly brain and the second part describes specific MR-appearances in some particular extrapyramidal diseases.

DESCRIPTIVE MR-CHANGES IN PATIENTS WITH EXTRAPYRAMIDAL DISEASES

Quantitative indices

Reported CT and MRI indices of brain atrophy in patients

with dementia overlap with indices of atrophy in healthy elderly subjects. That overlap can be reduced by controlling for diagnosis, age, and head size or by using volumetric rather than linear measures. The disadvantage of this approach is the time consuming computer program, however, such studies revealed that the severity of dementia in affected man was related to the extent of cerebral atrophy, measured as a reduced gray matter volume or as increased CSF spaces (Creasey et al 1986).

Periventricular phenomena

MRI is the most sensitive technique for detecting periventricular pathology (Gerard and Weisberg 1986). Abnormalities were seen in 30 % of the patients over age 60 (Bradley et al 1984).

Periventricular high intensity patterns on T2W I cannot be related only to normal aging. For the clinical significance of periventricular phenomena different patterns of increasing extent and intensity were delineated (Zimmermann et al 1986). While mild periventricular changes is a normal finding, more extensive lesions are associated with intracerebral pathology but the finding is often nonspecific. Periventricular white matter lesions are not necessarily pathognomonic of cerebrovascular disease.

White matter lesions

The white matter lucencies, termed leucoaraiosis, are defined as patchy or diffuse areas of decreased alternations on CT or abnormal signal intensity involving only the white matter and with no change in adjacent ventricles or sulci. White matter changes in MRI are related to aging to gradual changes in the chemical composition of the white matter with aging.

Leucoaraiosis appears to be a relatively common finding on MRI scans of elderly individuals and play a significant role in the development of intellectual impairment, however, the heterogeneous etiology of this condition must be taken into account. Subcortical and periventricular white matter lesions have been reported in completely asymptomatic patients without any history of TIA or stroke, risk factors or signs of dementia.

Thinning of corpus callosum

Thinning of corpus callosum was found in normal pressure hydrocephalus, obstructive hydrocephalus, and Alzheimer disease, but the significance of prevalence of this finding as well as the precise mechanism producing thinning of corpus callosum morphology are unknown (Jack et al 1987).

The CSF flow void sign

The CSF flow void sign is primarily due to the pulsatile nature of CSF flow rather than to bulk CSF flow. The CSF flow void sign was found in normal and abnormal conditions. At low field strength, the absence of a marked or moderate CSF flow void sign may militate against a diagnosis of normal pressure hydrocephalus (Jack et al 1987). The intrinsic sensitivity of MR to moving spins enable the visualization of CSF motion.

Iron deposition

On T2 weighted images at 1.5 Tesla low signal intensity is noted within ferritin containing structures as globus pallidus, red nucleus, substantia nigra, caudate nucleus and putamen. This phenomenon corresponds to physiologic and age related deposition of ferritin.

Drayer et al 1986 reported low intensity in the basal ganglia on T2WI which they attributed to iron deposition. Iron deposition is confirmed using Perl stains on autopsy specimens and the iron is presumed to be in the form of ferritin. Ferritin can be visualized after 6 months and generally increase until age 20 with a further increase noted after age 60.

Iron has also been noted in specific locations with certain disease states. Neuroaxonal dystrophy (Hallervorden Spatz disease) is characterized by marked ferritin deposition in the globus pallidus and the reticular portion of substantia nigra. The putamen and globus pallidus are involved in Parkinsons-disease and multisystem atrophy (Drayer 1986). In Huntington`s disease increased ferritin deposition have been found pathologically in the caudate nucleus and the putamen. On table 1 brain iron deposition is described in certain physiologic and pathologic states including the corresponding location (Fig. 1).

Brain Iron	Localization
Normal distribution	
infancy	absent
adult	GP/SN/RN/DN
aging	
Neuro-degenerative	
Parkinson`s disease	Putamen/SN compacta
Multisystem atrophy	Putamen/SN compacta
Chorea Huntington	Caudate, Putamen
Hallervorden Spatz	GP/SN reticulata
Alzheimer disease	Cerebral cortex
Demyelinating disease	Thalamus, Putamen
Neoplasm, Radionecrosis	
Hemorrhage	

Table 1
GP = globus pallidus, SN = substantia nigra, RN = red nucleus, DN = dentate nucleus

In the brain iron is taken up by dendrites and then transported along axons to the site of eventual utilization where it is stored by oligodendroglial cells. Diseases destroying the axons result in increased iron accumulation both near the lesion and at the point of dendritic uptake. Thus increased ferritin deposition has been noted in the region of demyelinating plaques in multiple sclerosis by pathological studies. In addition, periventricular processes that destroy axons may result in increased iron deposition in the thalami. However, the significance and specifity of increased iron deposition remain to be determined.

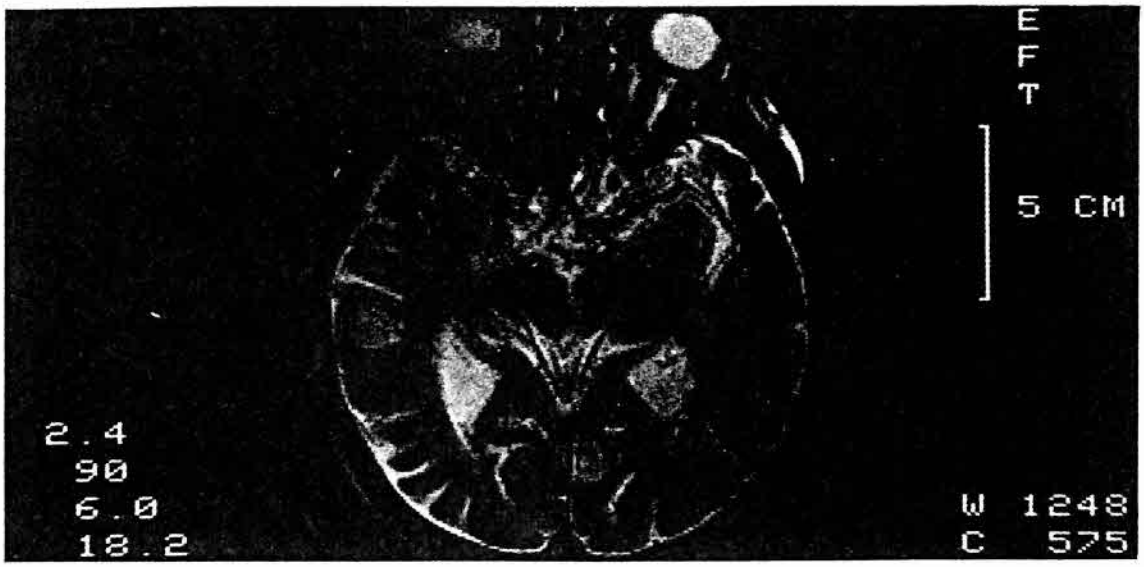

Fig. 1a: On T2-weighted image at 1.5 T low signal intensity is noted within ferritin containing structures. Axial section through midbrain demonstrates T2 shortening in red nucleus and substantia nigra.

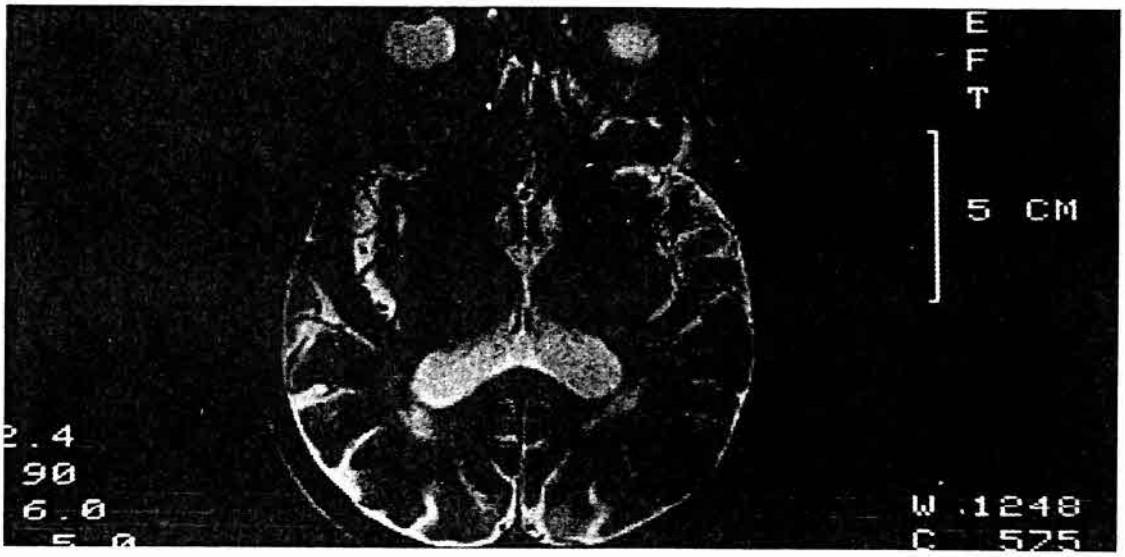

Fig. 1 b: Axial section through basal ganglia demonstrates T2 shortening in globus pallidus bilaterally.

MR-FINDINGS IN SPECIAL EXTRAPYRAMIDAL DISEASES

Parkinson`s disease

The first results of low and middle field MR in patients with Parkinson`s disease were rather disappointing even in cases with unilaterally accentuated lesions (Aichner et al 1985, 1986). Recently,several authors reported their results in Parkinson`s disease using 1.5 Tesla MR-machines (Drayer et al 1986, 1988; Pastakia et al 1986; Rutledge et al 1987). While the increased iron deposition into the basal ganglia seems to be a quite nonspecific finding, Rutledge et al described a restoration of signal in the substantia nigra as well as less specific forms of atrophy. Further studies are necessary to compensate the lack of correlation between chemically demonstrated iron distribution and metabolism.

Multisystem atrophy (MSA)

Neuropathologically, in MSA neuronal loss and/or gliosis is present in a number of central nervous system areas: substantia nigra, locus ceruleus, caudate, putamen, cerebellar cortex, pontine nuclei, inferior olives, dorsal vagal nucleus, anterior and lateral horns of the spinal cord and pyramidal tract. There are relatively few MR-examinations of patients with MSA. MR imaging at a field strength of 1.5 T permits the in vivo mapping of brain iron and thereby provides the monitoring of normal or pathologic aging. In patients with multisystem atrophy with features of autonomic failure (Shy-Drager-syndrome), striatonigral degeneration, olivopontocerebellar atrophy (OPCA) and progressive supranuclear palsy (PSP) Drayer reported 1986 a prominent topographically specific decrease in T2 in 6 patients with MSA. MRI in a case of Shy-Drager-syndrome revealed a loss of signal from the putamen which may prove not be specific for Shy-Drager-syndrome (Pastakia et al 1986).

In our study of 9 patients examined by 1.5 T no specific finding could obtained. Table 2 lists all clinical data. Seven patients showed cerebral atrophy while 5 patients demonstrated cerebellar atrophy which seemed to be more specific for MSA than brain atrophy. No change in parenchymal signal was noted. Ferritin deposition was only found in two patients with MSA. The most prominent result of our MSA-study was the marked cerebellar atrophy. Further studies using 3-D techniques as well as quantitative measurements are necessary to improve the visualization of the neuropathologically wellknown changes within the different areas of the central nervous system.

INITIAL	AR	T	OPTOMOTORIC DYSFUNCTION	ATAXIA	DYS-ARTHRIA	DYS-PHAGIA	BLADDER DYSF.	HYPOTONIA
K.J.	++	-	+	++	++	++	+	++
K.M.	+	-	++	++	++	++	-	++
P.I.	++	±	+	++	+	+	+	+
G.A.	++	-	+	++	+	+	+	++
K.H.	++	-	±	++	++	+	+	++
H.M.	++	-	+	+	++	++	+	++
G.E.	++	-	+	±	+	-	-	+
F.L.	+	-	++	++	++	++	-	++
G.L.	++	-	+	++	+	±	-	+

INITIAL	SEX	AGE	LENGTH OF ILLNESS a	DIAGNOSIS-LABEL	DEMENTIA	MRI-FINDINGS
K.J.	m	59	23	OPCA	+	CEREBELLAR ATROPHY FERRITIN DEPOSITION
K.M.	f	51	5	PSP	+	BRAIN ATROPHY
P.I.	f	67	3	OPCA	+	BRAIN ATROPHY
G.A.	f	61	3	SHY DRAGER	-	CEREBELLAR ATROPHY
K.H.	f	65	3	OPCA	-	CEREBELLAR AND CEREBRAL ATROPHY
H.M.	f	50	5	OPCA	-	CEREBELLAR AND CEREBRAL ATROPHY
G.E.	f	68	8	?OPCA	+	CEREBRAL ATROPHY FERRITIN DEPOSITION
F.L.	f	68	6	OPCA	+	CEREBELLAR AND CEREBRAL ATROPHY
G.L.	f	66	4	OPCA	+	CEREBRAL ATROPHY

Table 2: Clinical and MR data of 9 patients with MSA.
AR = akinetic-rigid syndrome, T = tremor.

To improve the sensitivity and specifity of MRI in diagnosing and monitoring MSA the development of 3-D imaging with quantitative and volumetric measurements has to be strengthened. Up to now, most of the MRI results in Parkinsonism and MSA remain unspecific (Fig. 2).

Fig. 2a: Sagittal FLASH sequence of a patient with the clinical diagnosis of OPCA demonstrates marked cerebellar atrophy.

Fig. 2b: Axial T2 weighted image through the cerebellum shows moderate cerebellar atrophy.

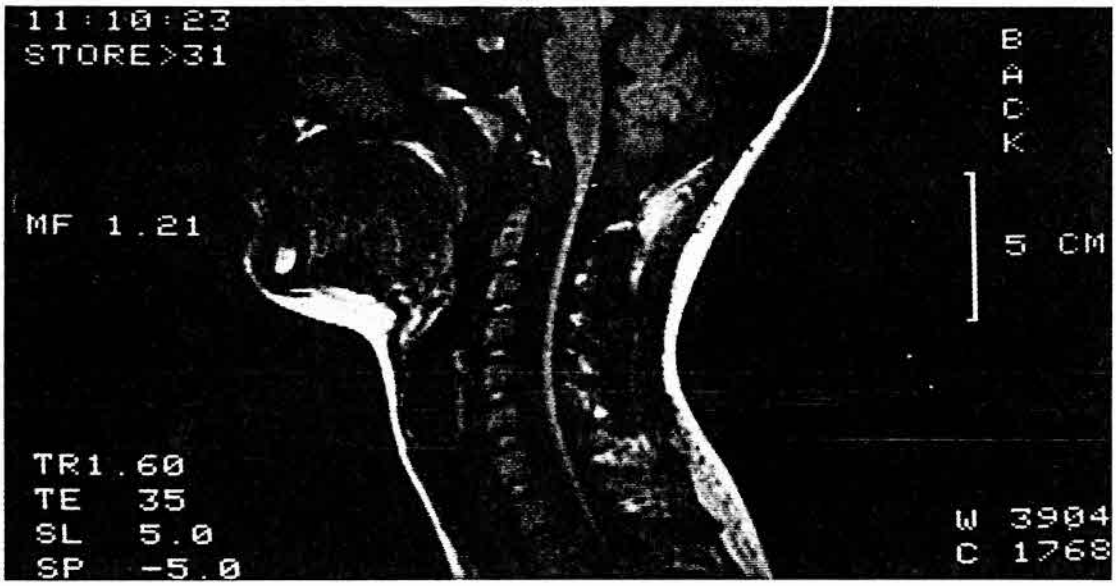

Fig. 2c: Sagittal section, proton density weighted, through the cervical spine demonstrates spinal cord atrophy in a patient with spinocerebellar degeneration.

Chorea Huntington

Little experience exist in highfield MRI of Huntington disease. Simmonds et al 1986 reported on 4 patients, the same group published 1987 MR findings in 6 patients with Huntington disease. They described mild to moderate cerebral atrophy, flattening of the caudate head and uniformly thick periventricular hyperintensity.

Dystonia

Since primary dystonia have no known pathology MRI, up to now, has not shown any abnormality.Secondary dystonia, which are associated with metabolic diseases, have several known pathologic changes (Rutledge 1987). Wilson`s disease is an example demonstrating lesions in the putamen and caudate as well as in the midbrain, pons, and cerebellum (Starosta-Rubinstein 1987).

Essential blepharospasm (EB)

MRI studies were performed in 7 patients with EB from whose 5 patients were classified as Meige-syndrome. Table 3 shows clinical data. In one case a meningeoma was seen accidentally and another case showed white matter lesion, however, MRI did not show a EB related lesion.

INITIAL SEX	AGE	PERIOD OF DISEASE[a]	BSP	OMD	ET	MEIGE SYNDROME	BTX	MRI FINDINGS
K.H. f	51	3	++	++	-	+	+	MENINGEOMA
H.K. f	63	3	++	++	-	+	+	NORMAL
L.A. f	66	5	++	+	-	+	+	NORMAL
T.F. f	68	5	++	-	+	-	+	NORMAL
K.E. f	62	8	++	+	-	-	-	NORMAL
K.G. f	78	1	++	-	-	-	-	NORMAL
A.G. f	77	4	++	+	-	+	+	WML

Table 3: Clinical and MR-data of 7 patients with essential blepharospasm. BSP = blepharospasm, OMD = oro-mandibular dystonia, ET = essential tremor, BTX = Botulinum Toxin, WML = white matter lesion.

CONCLUSION

Previous reports studying patients with extrapyramidal diseases showed no evidence of any specific findings. T2 weighted imaging of the brain at 1.5 Tesla results in excellent display of basal ganglionic structures. MRI easily show local or more generalized atrophy, increased or decreased signal intensity, changes in the content of iron within the extrapyramidal nuclei as well as periventricular and deep white matter abnormalities occuring under physio-

logic or pathologic conditions of the elderly population. Further studies will be required to assess whether MR can detect additional findings, particularly, when 3-D imaging and quantitative measurement will be used for.

REFERENCES

Aichner F, Poewe W, Birbamer G, Gerstenbrand F (1986). Erfahrungen mit dem Magnetic Resonance Imaging,beim Parkinsonsyndrom. Roche Wiss. Dienst. Hrsg: Schnaberth G, Auff E. 385 - 388

Aichner F, Willeit J (1986). Magnetic resonance imaging of the central nervous system. In: PET and NMR: new perspectives in neuroimaging and clinical neurochemistry Ed: Battistin L, Gerstenbrand F. A.R. Liss. Inc. New York, 381 - 406

Bradley WG, Waluch V, Wycoff RR (1984). Differential-diagnosis of periventricular abnormalities in MRI of the brain. Noninvasive Medical Imaging 1: 35 - 41

Creasey H, Schwartz M, Frederickson H, Haxby JV, Rapoport STI (1986). Quantitative computed tomography in dementia of the Alzheimer type. Neurology, 36: 1563 - 1568

Drayer BP, Olanow W, Burger P, Johnson GA, Herfkens R, Riederer S (1986). Parkinson Plus Syndrome: diagnosis using high field MR Imaging of brain iron. Radiol. 159: 493 - 498

Drayer BP, Burger P, Darwin R, Riederer S, Herfkens R, Johnson GA (1986). Magnetic resonance imaging of brain iron. AJNR 7: 373 - 380

Drayer BP (1988). Imaging of the aging brain. Part II Pathologic conditions. Radiol 166: 797 - 806

Gerard G, Weisberg LA (1986). MRI periventricular lesions in adults. Neurology, 36: 998 - 1001

Jack CR, Mokri B, Laws ER, Houser OW, Baker HL, Peterson RC (1987). MR findings in normal pressure hydrocephalus: significance and comparison with other forms of dementia. J.Comp.Assist: Tomogr. 11: 923 - 931

Pastakia B, Polinsky R, Di Chiro G, Simmons JT, Brown R, Wener L (1986). Multiple System Atrophy (Shy-Drager Syndrome): MR imaging. Radiol. 159: 499 - 502

Rutledge JN, Hilal SK, Silver AJ, Defendini R, Fahn S (1987). Study of movement disorders and brain iron by MR. AJR 149: 365 - 379

Simmons JT, Pastakia B, Chase TN, Shults CW (1986). Magnetic resonance imaging in Huntington disease. AJNR 7: 25 - 28

Starosta-Rubinstein S, Young AB, Kluin K, Hill G, Aisen AM, Gabrielsen T, Brewer GJ (1987). Clinical assessment of 31 patients with Wilson`s disease. Arch. Neurol. 44: 365 - 370

Zimmermann RD, Fleming CA, Lee BCP, Saint Louis LA, Deck MDF (1986). Periventricular hyperintensity as seen by Magnetic resonance: prevalence and significance. AJNR 7: 13 - 20

Aichner F, Feiber S, Poewe W, Ransmayr G, Gerstenbrand F.

MRI of extrapyramidal Diseases

In:

Aging Brain and Dementia: New Trends in Diagnosis and Therapy

by Leontino Battistin (Editor), Franz Gerstenbrand (Editor)

The aging of the brain represents the most important problem among those related to aging in general, and this book offers a full account of the processes occurring in both the normal aging and pathological brain. Using a multidisciplinary approach, Aging Brain and Dementia covers biological and clinical investigations of brain function and examines in detail epidemiological, clinical, biochemical, psychological, diagnostic, and therapeutic aspects of aging brain and dementia. The book includes findings that augment existing knowledge of when an impairment of metabolism and neurotransmission is still functional, and therefore reversible, and when it becomes morphological and irreversible. In addition, it shows how clinical advances lead to the successful evaluation of cognitive abilities and disorders of aged patients as well as those with dementia. Read Less

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