Magnetic resonance imaging in progressive supranuclear palsy and other parkinsonian disorders

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Summary. High field intensity MRI may demonstrate signal abnormalities consistent with deposits of iron or other paramagnetic substances in several extrapyramidal disorders. Hallervorden-Spatz disease was the only disorder widely known to have iron deposits in the pallidum, that are now easily demonstrated in vivo by MRI. However, lower field intensity MRI may also demonstrate characteristic findings.

In progressive supranuclear palsy, definite atrophy of the midbrain and of the region around the third ventricle is seen in slightly more than half of the cases. Minimal signal abnormalities are sometimes seen in the periaqueductal region, but MRI studies remain of little help in establishing the diagnosis of the disease.

Asymmetric atrophy in the parietal regions is seen in corticobasal degeneration, as expected from pathological studies. Minimal alterations may be seen in the substantia nigra in Parkinson's disease.

The most interesting MRI findings are observed in multiple system atrophies. Variable abnormal signal intensities, depending on the field intensity, are visible in the putamen in striatonigral degeneration and in Shy-Drager syndrome; in this latter condition the abnormalities are due to its striatonigral degeneration component. Atrophy of the pons, middle cerebellar peduncles, and cerebellum, and signal abnormalities in a characteristic distribution are visible in olivopontocerebellar atrophy.

A combination of these posterior fossa abnormalities and putaminal alterations may confirm the involvement of the cerebellar and extrapyramidal systems in multiple system atrophies.

The interest in Magnetic Resonance Imaging (MRI) of parkinsonian disorders arose about six years ago, when it was recognized that high field intensity MRI could demonstrate iron in the brain (Drayer et al., 1986a) and that some parkinsonian syndromes presented abnormal distribution of iron or other paramagnetic substances in the basal ganglia (Drayer et al., 1986b; Pastakia et al., 1986).

Iron is not present in the brain at birth, but accumulates during life with an uneven distribution; it is present in greater amounts in the basal ganglia,

particularly in the pallidum, in the substantia nigra, red nucleus and dentate nucleus. Iron or other paramagnetic elements cause a shortening of T2 relaxation time; therefore, at high field intensity MRI, the areas where iron accumulates present a low signal intensity in T2-weighted images (Drayer et al., 1986b; Gomori et al., 1985). The distribution of this low signal intensity correlates exactly with the intensity of the blue coloration of the brain at the Perls' staining for iron (Drayer et al., 1986a).

The extrapyramidal disorder which for years has been known to present abnormal accumulation of iron in the pallidum is Hallervorden-Spatz disease. A remarkable loss of signal intensity in the pallidum in T2-weighted images was expected, therefore, in this disease; indeed, the first reports of MRI findings in Hallervorden-Spatz disease confirmed this expectation, but also demonstrated the presence of a small area of high signal intensity in the anteromedial part of this nucleus (Rutledge et al., 1987; Sethi et al., 1988). This was called by Sethi et al. the "eye-of-the-tiger sign" (Sethi et al., 1988). The significance of this high signal intensity area was unclear from the previous pathological reports; only Dooling et al. had mentioned that destructive changes and gliosis were more evident in the internal segment of the pallidum (Dooling et al., 1974). Recently, we could compare the MRI findings of our eight clinical cases of Hallervorden-Spatz disease, which all presented the "eye-of-the-tiger sign", with the pathological specimens of two proven cases of Hallervorden-Spatz disease examined by Halliday in Winnipeg (Canada). The area of high signal intensity corresponds to an area of "loose" tissue, with vacuolization and less iron than found in the rest of the pallidum, where the tissue is more "dense" and contains greater amounts of iron (Savoiardo et al., 1993).

An additional observation we could make was that when we examined our cases of Hallervorden-Spatz disease with intermediate field intensity MRI (i.e. 0.5 Tesla (T) rather than 1.5 T) we could easily demonstrate the area of high signal intensity, while the loss of signal intensity due to iron was poorly detectable; with gradient echo images, however, iron was detected or better demonstrated also at 0.5 T (Fig. 1).

Therefore, when discussing the magnetic susceptibility effects of iron or other paramagnetic substances, it is important to make clear whether intermediate or high field intensity MRI is used, and whether spin echo or gradient echo techniques, which are more sensitive to the magnetic susceptibility effects, are employed.

We shall now discuss the MRI findings of progressive supranuclear palsy, which is the main topic of this discussion, and then of other parkinsonian disorders, mainly of multiple system atrophies.

Progressive supranuclear palsy or Steele-Richardson-Olszewski syndrome

From pathological reports, progressive supranuclear palsy (PSP) is known to be associated with atrophy of the midbrain (Barr, 1979). Neuroradiological studies, therefore, may support the diagnosis by demonstrating this

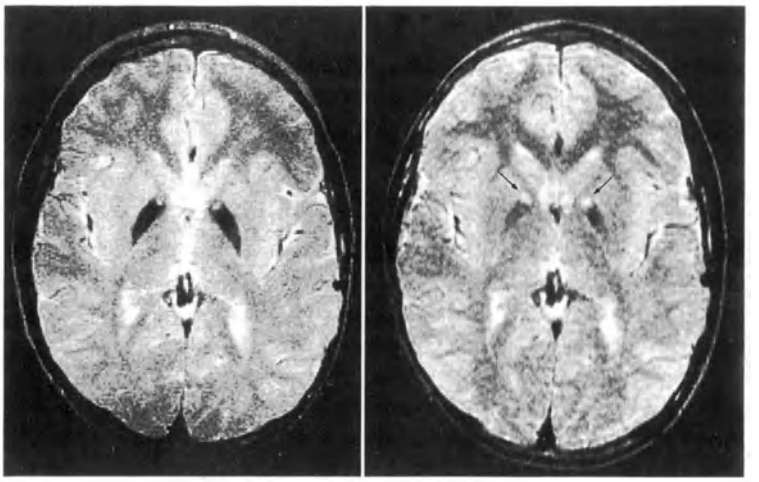

Fig. 1. Hallervorden-Spatz disease. T2-weighted images obtained at different field intensities (**A** 1.5 T; **B** 0.5 T) demonstrate the dependence of the magnetic susceptibility effects due to iron in the pallidum on the field strength; the loss of signal intensity caused by iron is much more evident at 1.5 T (**A**). The area of high signal intensity in the anteromedial part of the pallidum (eye-of-the-tiger sign) is more evident at 0.5 T (arrows, **B**). [From AJNR (Savoiardo et al., 1993), with permission]

atrophy; this demonstration was obtained first by pneumoencephalography (Bentson and Keesey, 1974) and then by computed tomography (CT) (Masucci et al., 1985; Schonfeld et al., 1987). The first MRI report included PSP among the multiple system atrophies (MSAs) and suggested that abnormal distribution of iron in the lentiform nucleus (with predominant loss of signal intensity in T2-weighted images at high field intensity in the putamen rather than in the pallidum) was the characteristic feature of these diseases (Drayer et al., 1986b). We could not confirm these findings in PSP (Savoiardo et al., 1989). We subsequently expanded our observations to 20 cases and the relevant findings will be reported here.

The 20 patients with clinical diagnosis of PSP were followed by a group of neurologists with particular experience in extrapyramidal disorders. There were 11 males and 9 females; age ranged from 56 to 79 years (mean 64.6 years). Length of the disease ranged from 2 to 10 years (mean 3.8 years).

The MRI studies were performed with a 0.5 T equipment in 10 cases and with 1.5 T machine in 13 cases. Three patients were therefore examined at both field strengths.

The MRI findings were evaluated by three experienced neuroradiologists, but measurements of the size of the brainstem or other structures were not used.

The expected finding of atrophy of the midbrain was unquestionable in 11 cases; in the other 9 patients the size of the midbrain was borderline or appeared normal. When atrophy was present, it was recognizable both in

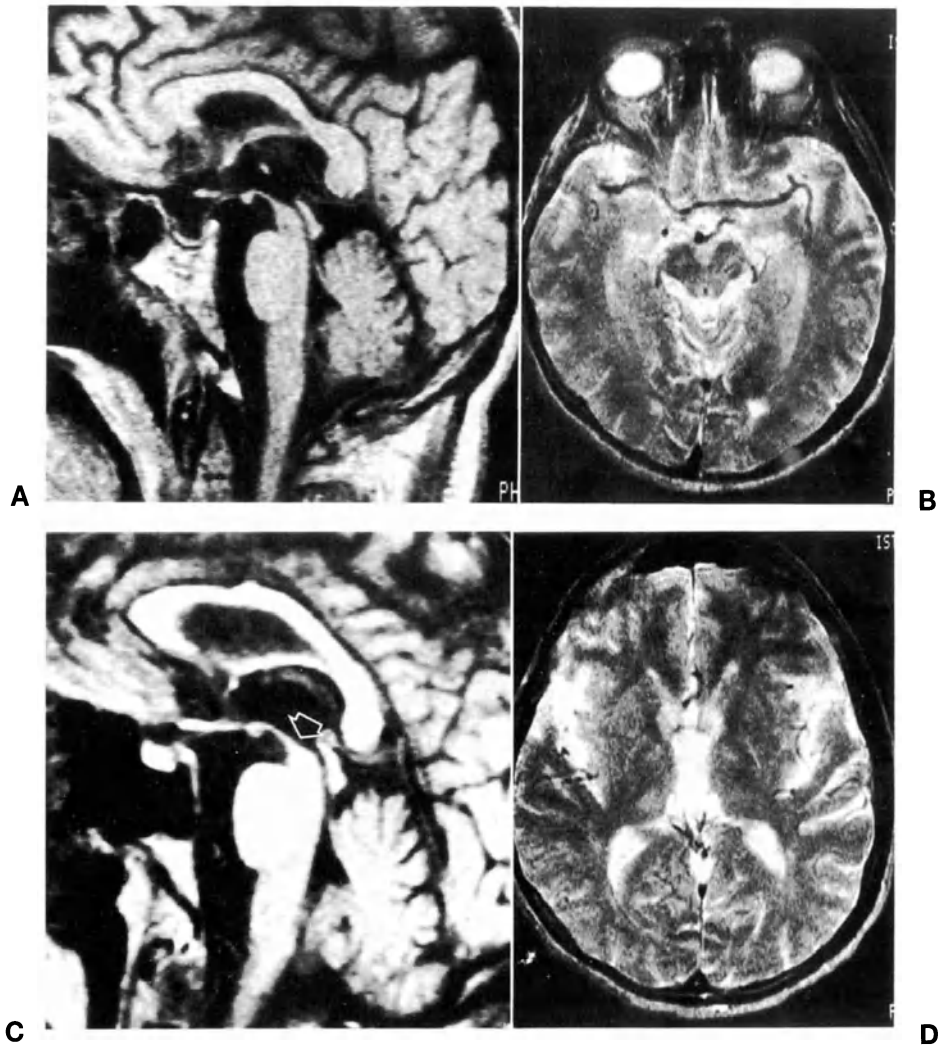

Fig. 2. Progressive supranuclear palsy. Sagittal T1-weighted images demonstrate definite midbrain atrophy in about half of the cases (**A**); atrophy of the dorsal midbrain is also recognizable in axial sections (**B** T2-weighted image). Hyperextension of the head may be present (**C**); atrophy involving the brain tissue around the third ventricle also causes an inferior convexity of the floor of the third ventricle (open arrow, **C** sagittal T1-weighted section), and a disproportionate enlargement of the third ventricle compared to the lateral ventricles (**D** axial T2-weighted image)

transverse and in sagittal sections. Sagittal sections were particularly helpful in demonstrating thinning of the quadrigeminal plate, more marked in its superior part (Fig. 2A and B).

Another atrophic feature consisted in dilatation of the third ventricle, which was disproportionately wider than the lateral ventricles in 11 patients; 10 of them coincided with the patients who also had definite midbrain atrophy. The widening of the third ventricle and the midbrain atrophy often

determined a concave aspect of the posterior part of the floor of the third ventricle on the midline sagittal section (Fig. 2C and D).

Occasionally one could guess the diagnosis of PSP merely observing the first sagittal sections, when attention was called by a hyperextended position of the head and the midbrain atrophy was then recognized (Fig. 2C).

Regarding signal abnormalities, the findings were also subtle and inconstant. The most common signal abnormality observed in our series of PSP was a slight increase in signal intensity in intermediate or proton density images in the pariaqueductal region (Fig. 3A and B), where gliosis is found in pathologic specimens. No definite abnormalities could be recognized in this region in T2-weighted images, which sometimes demonstrated a prominent loss of signal intensity in the region of the substantia nigra, with smudging of its margin toward the red nucleus at high field intensity MRI. Of the 13 cases examined with high field intensity MRI, 11 presented normal distribution of iron in the lentiform nucleus, i.e. low signal intensity in T2-weighted images in the pallidum and normal signal or minor loss of signal intensity in the putamen (Fig. 3C). In 2 cases, the loss of signal intensity of the putamen equalled or was perhaps superior to that seen in the pallidum; however, one was a patient of 79 years of age, and it has been reported that in old age the accumulation of iron in the putamen may equal that in the pallidum. Therefore, only one patient of age 66 is left with abnormal distribution of signal intensity in the basal ganglia, a finding that remains distinctive of MSAs.

We had the chance of examining with 1.5 T MRI a 1 cm thick coronal section through the basal ganglia in a case of pathologically proven PSP; the T2-weighted images did not exhibit abnormalities of signal intensity consistent with abnormal iron deposition, a finding which was confirmed by the normal Perls' staining.

A rare signal abnormality observed in 3 of the 13 cases examined at 1.5 T was low signal intensity in T2-weighted images in the superior colliculi which did not appear atrophic (Savoiardo et al., 1989).

Finally, several patients had diffuse supratentorial atrophy with widening of cisterns and sulci, and some of them had rare areas of signal abnormalities in the white matter of the cerebral hemispheres, mostly in the periventricular region. However, these changes were considered not in excess to what is usually seen in a group of subjects of the same age.

In conclusion, all the significant abnormalities that correlate with the known pathological findings of PSP were seen in the region of the midbrain and around the third ventricle. However, definite although subtle abnormalities were seen in only about half of the patients with clinical diagnosis of PSP and, therefore, MRI remains of little help in establishing the diagnosis of this disease.

Corticobasal degeneration

Corticobasal degeneration (CBD) is a recently rediscovered disease, for which the importance of imaging studies has not yet been determined. From

Fig. 3. Progressive supranuclear palsy. Minimal signal hyperintensity may be observed only in intermediate or proton density images in the periaqueductal region (**A** axial section, arrowheads; **B** sagittal section, arrow, in two different patients). The signal intensity in the lentiform nucleus is normal; low signal intensity consistent with abnormal putaminal deposits of iron is not observed (**C** T2-weighted axial section in a 66-year-old man with PSP, 1.5 T)

pathological studies diffuse histological changes in the substantia nigra, midbrain tegmentum, subthalamic and lentiform nuclei are known to occur in association with parietal or frontoparietal atrophy on the side opposite to the alien hand, which is a characteristic clinical feature (Gibb et al., 1989; Rebeiz et al., 1968).

There are clinical similarities with PSP, in terms of axial rigidity, supranuclear gaze palsy, and bradykinesia (Gibb et al., 1989).

With MRI we examined six patients with clinical diagnosis of CBD, 4 males and 2 females. Their age ranged from 63 to 76 years (mean 67.5 years). In only one patient was the midbrain considered to present borderline atrophy. In all the other cases the midbrain presented normal size. Diffuse supratentorial atrophy was observed in all the cases but predominance in one parietal region was mentioned only in two cases on the first reading. However, after being alerted of possible asymmetries, we reviewed the cases and could clearly identify a greater degree of parietal atrophy on the side opposite to the dystonic alien hand in all six cases, without knowing which side should have been affected. Awareness of the disease is therefore essential for noticing the focal atrophy (Fig. 4). No signal abnormalities were seen except for a minimal increase in signal intensity in the affected parietal cortex in one case (Fig. 4C), and for a few lacunes in the basal ganglia in two other cases.

Parkinson's disease

MRI studies on Parkinson's disease have been focussed on the changes in signal intensity in the substantia nigra. High field MRI may demonstrate low signal intensity in this region with smudging of its posterior border toward the red nucleus (Braffman et al., 1988; Duguid et al., 1986). The anterior extension of the low signal intensity usually includes the medial part of the cerebral peduncle. Therefore, the assessment that the hypointensity is the expression of iron deposition in the pars reticulata and that the reduction of the normal signal intensity between substantia nigra and red nucleus is due to atrophy of the pars compacta seems hazardous. To our knowledge, there are no studies that correlate the MRI findings with the histological demonstration of iron deposition in this region.

Another study on Parkinson's disease emphasized the "restoration" of normal signal intensity in T2-weighted images in the dorsolateral part of the substantia nigra (Rutledge et al., 1987) (Fig. 5).

These two signs may coexist, but their importance in supporting the diagnosis of Parkinson's disease or other parkinsonian disorders is modest. In patients with parkinsonian disorders poorly responding to therapy, it seems more important to look for changes in the putamen (see the following section on MSAs) that may indicate a poor prognosis with evolution toward a multiple system atrophy.

Multiple system atrophies

When we started to study patients with MSAs, we decided to try to separate patients who had prominent signs of autonomic failure, accompanied by cerebellar and parkinsonian signs of variable severity, from patients who presented predominant extrapyramidal signs not responding to therapy, and from patients who presented mainly a cerebellar disorder with no or mild

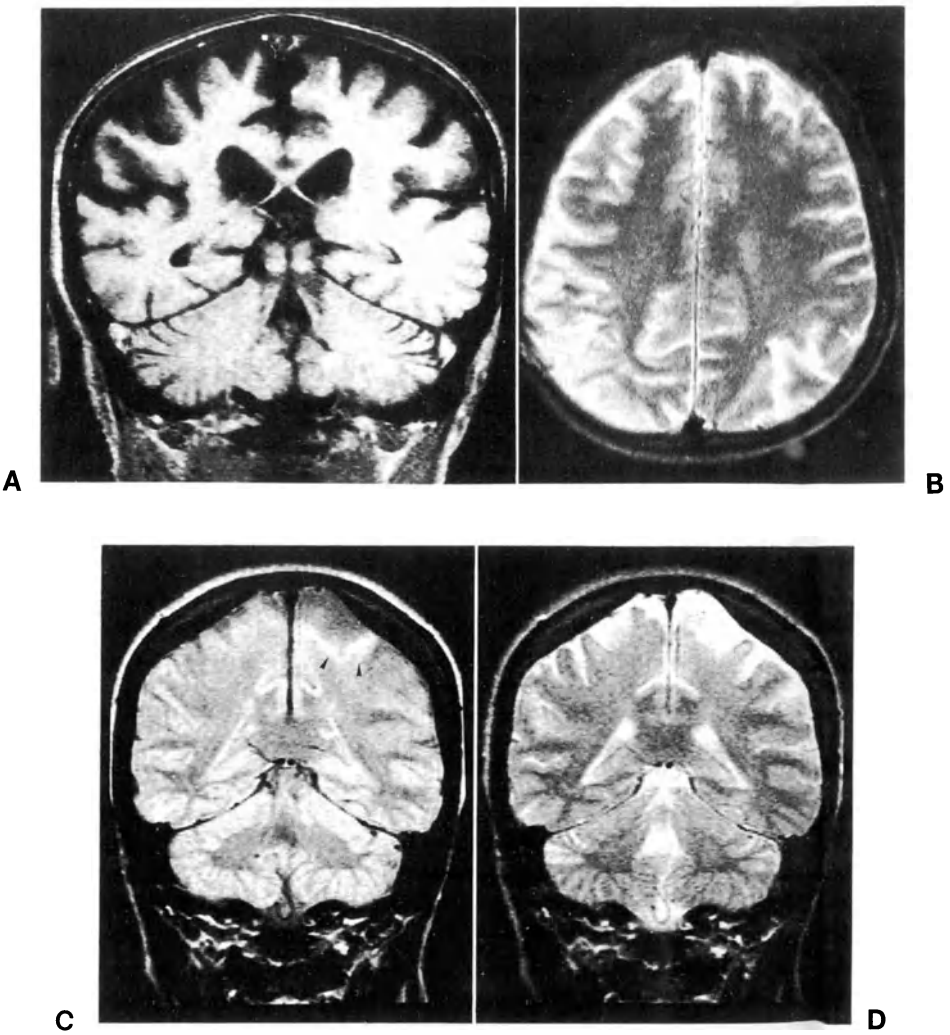

Fig. 4. Corticobasal degeneration. Coronal T1- (**A**) and axial T2-weighted images (**B**) in the same patient demonstrate right parietal atrophy; the patient had a left alien hand. Minimal high signal intensity in the left parietal atrophic cortex was seen only in one patient in proton density images (**C** arrowheads); no abnormal signal intensity is recognizable in the T2-weighted image (**D**). This patient had a right alien hand sign

extrapyramidal and autonomic disturbances. In other terms, we tried to separate patients with *Shy-Drager syndrome (SDS)* from patients with *striatonigral degeneration (SND)* and with *olivopontocerebellar atrophy (OPCA)*. This separation was aimed at finding the characteristic MRI features of these different entities, in order to subsequently verify whether patients who presented with marked involvement of all the three systems, i.e. extrapyramidal, autonomic, and cerebellar, could also be recognized by imaging studies as having multiple systems involvement.

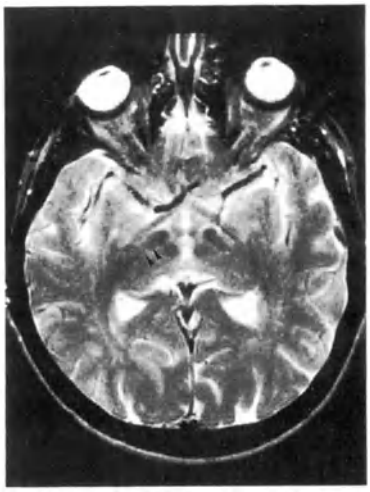

Fig. 5. Parkinson disease. In this patient with left hemiparkinson a slight asymmetry of signal suggests restoration of signal intensity in the lateral part of the substantia nigra on the right side (arrowheads, T2-weighted image, 1.5 T)

After collecting several cases of SDS, SND and OPCA, we realized that there was identity of MRI findings in patients with SDS and SND, while patients with OPCA presented abnormalities confined to the posterior fossa, that corresponded exactly to the distribution of gross and microscopic abnormalities described in the pathology of this disease. There were also cases, indeed, with marked clinical involvement of the three systems, which presented, on MRI, combination of abnormalities in posterior fossa, as seen in OPCA, and in the putamen, as seen in SDS and SND.

We now have seen more than 60 cases with MSA and, although an occasional patient clinically diagnosed as an MSA case may present normal or questionable MRI study, we found a quite reliable correspondence between clinical and MRI findings, particularly in OPCA. In our Institute, a tentative clinical diagnosis of OPCA in a sporadic case is now accepted only if it is supported by the results of MRI studies.

Striatonigral degeneration and Shy-Drager syndrome

SDS is considered a synonym of MSA by many authors (Oppenheimer, 1984; Quinn, 1989). Here we considered as SDS cases the patients who had prominent dysautonomic signs, with minimal cerebellar and extrapyramidal involvement. They were separated from patients with marked involvement of all the three systems, who were more clearly MSA patients.

We have already said that the MRI findings observed in SND and SDS are identical. In fact, SND is an associated or constitutive finding in a high proportion of cases of SDS (Oppenheimer, 1984). MRI, so far, has not

demonstrated abnormalities in the spinal cord, where the abnormalities responsible for the autonomic failure should be sought for in the intermediolateral columns. Patients with pure autonomic failure studied with MRI of the brain did not exhibit any abnormalities (Brown et al., 1987).

The abnormalities seen in brain MRI of SND and SDS at 1.5 T are different from those seen in 0.5 T studies, but in both are confined to the putamen.

In 1.5 T studies, there is low signal intensity in T2-weighted images in the putamen, which is more marked than in the pallidum, with an inversion, therefore, of the normal signal distribution within the lentiform nucleus (Fig. 6). This putaminal low signal intensity is consistent with increased deposits of iron, but other paramagnetic substances, such as manganese, neuromelanin and "hematin" pigments (Borit et al., 1975; Dexter et al., 1991; Pastakia et al., 1986), can also be responsible for or contribute to this finding.

In the most lateral and posterior part of the putamen a thin rim of hyperintensity bordering the hypointensity is sometimes visible in T2-weighted images (Fig. 6).

The substantia nigra and the red nucleus sometimes presented a lower signal intensity than in normal subjects, but the low signal intensity in the putamen remained the most reliable and easily observable abnormality.

In 0.5 T studies, the low signal intensity in T2-weighted images due to iron or other paramagnetic substances is barely evident or absent. On the contrary, the MRI hallmark of these diseases at 0.5 T is putaminal hyperintensity in proton density and T2-weighted images, even in cases in which hyperintensity is not recognizable at 1.5 T. In cases with a thin band of hyperintensity on 1.5 T studies, 0.5 T examinations show a more evident and larger putaminal hyperintensity in T2-weighted images (Savoiardo et al., 1989, 1990) (Fig. 7A and B).

In our series, low signal intensity in the putamen at 0.5 T was well seen only in 3 of 10 SND cases (Fig. 7C), while was not seen in the 9 SDS cases. Although we did not find any difference between SND and SDS in 1.5 T studies, it is possible that the detection of iron at 0.5 T in some SND cases may indicate a greater amount of iron and a greater putaminal involvement in SND compared with SDS cases. This may correlate with the more severe extrapyramidal involvement of the patients with SND than of the patients with SDS.

The different findings obtained by 1.5 and 0.5 T studies are not contradictory; in 0.5 T examinations, the high signal intensity consistent with increased amounts of water, likely related to gliosis, neuronal loss, and spongiosis, is not masked by the presence of paramagnetic substances, which cause a signal loss in T2-weighted images proportional to the square of the magnetic field; therefore, at 1.5 T, these substances exert a magnetic susceptibility effect 9 times greater than at 0.5 T (Gomori et al., 1985; Savoiardo et al., 1989, 1990).

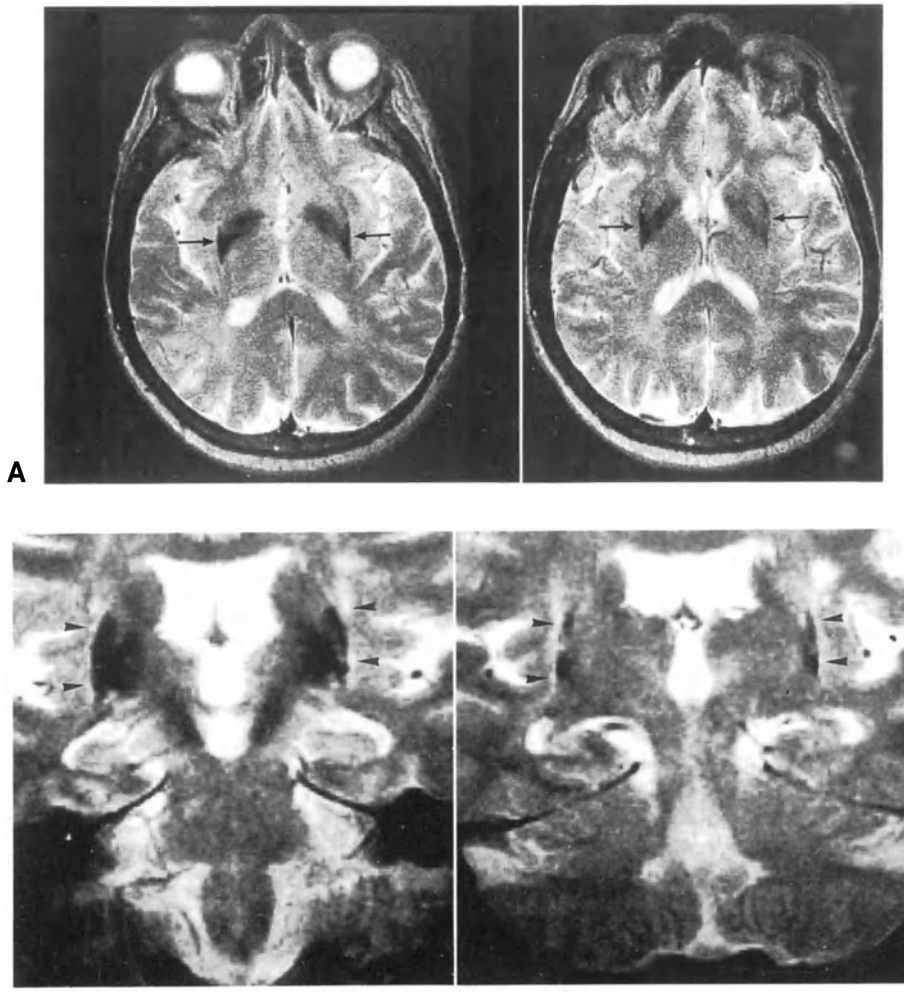

Fig. 6. Multiple system atrophies. At 1.5 T, involvement of the putamen is manifested by very low signal intensity in T2-weighted images. The loss of signal intensity of the putamen (arrows) prevails over that of the pallidum with reversal of the normal distribution (**A** and **B** axial sections; compare with Fig. 3C). This patient had severe involvement of extrapyramidal, autonomic and cerebellar systems and presented MRI abnormalities also in posterior fossa. Similar findings are present in a patient with diagnosis of Shy-Drager syndrome, who did not present clinical nor MRI findings of OPCA. Also note thin band of lateral hyperintensity (arrowheads; **C** and **D** coronal sections). [From J Comput Assist Tomogr (Savoiardo et al., 1989), with permission]

Olivopontocerebellar atrophy

OPCA may be diagnosed by the pathologist immediately at the inspection of the brain, when he observes atrophy of the pons, middle cerebellar peduncles, and cerebellum, usually more marked in the hemispheres than in the vermis. The diagnosis is then confirmed by the histological examination

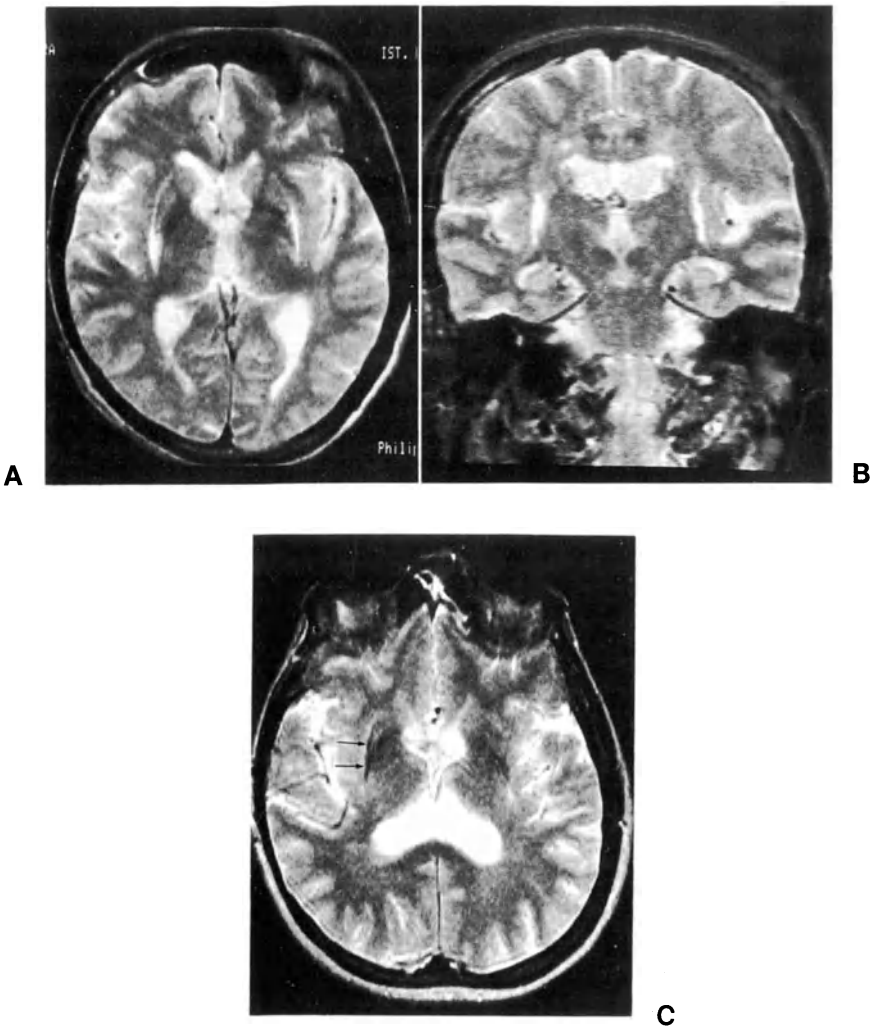

Fig. 7. Multiple system atrophies. In 0.5 T studies, high signal intensity consistent with increased amounts of water is observed (**A** axial, and **B** coronal T2-weighted sections in a case of Shy-Drager syndrome; same patient as in Fig. 6C and D. [From J Comput Assist Tomogr (Savoiardo et al., 1989), with permission.] In a case of striatonigral degeneration, low signal intensity particularly in the right putamen is present (**C** axial T2-weighted section, arrows)

of these structures. There is loss of cells in the pontine nuclei and degeneration of their fibers, which course as transverse pontine fibers in the anterior and posterior part of the basis pontis crossing on the midline in the raphe, running then in the middle cerebellar peduncles to reach the cerebellum. In the cerebellum there is a variable degree of degeneration of Purkinje cells; their fibers to the dentate nuclei also degenerate and the cerebellum

becomes gliotic. However, since the cells of the dentate nuclei do not degenerate, their fibers which run in the superior cerebellar peduncles to reach the red nuclei and the thalami remain intact. A retrograde cell loss of the inferior olives, secondary to cortical cerebellar lesions, is also seen (Oppenheimer, 1984). With MRI studies, the same findings can be observed. The distribution of atrophy is particularly well seen in T1-weighted images in sagittal and coronal sections. In midline sagittal sections, the normal bulge of the pons above the profile of the medulla oblongata diminishes, due to flattening of its inferior part. In more advanced cases, the whole pons is atrophic. The fourth ventricle enlarges and the cerebellar vermis becomes atrophic, with widened sulci (Fig. 8A and B). The coronal sections, however, demonstrate that atrophy involves equally or in a more severe degree the cerebellar hemispheres (Fig. 8C). These sections also demonstrate the atrophy of the middle cerebellar peduncles. Normally, a coronal section through the central or posterior part of the pons shows the rounded contour of the middle cerebellar peduncles; in OPCA, this contour becomes pointed (Fig. 8D). Obviously, these atrophic changes are also recognizable in axial and sagittal sections. No signal abnormalities are recognizable in T1-weighted images (Savoiardo et al., 1990).

In intermediate or proton density and T2-weighted images, slight signal hyperintensity is seen in the degenerated areas. Therefore, signal hyperintensity is seen in the transverse pontine fibers, in the middle cerebellar peduncles, and in the whole cerebellum (Fig. 9). Very rarely mild signal abnormalities are also recognizable in the inferior olives. The fibers that do not degenerate maintain normal signal intensity. Therefore, the pyramidal tracts, the superior and inferior cerebellar peduncles become well identifiable because they stand out by their normal signal intensity against the abnormal background (Savoiardo et al., 1990) (Fig. 9A, C and D). The most convincing demonstration of the distribution of signal abnormalities is obtained by comparing the proton density and T2-weighted images with the histological sections stained for myelin (Fig. 10, compare with Fig. 9A and C).

In axial sections, sometimes, the signal abnormalities of the cerebellum are poorly appreciated, particularly if they are very mild, because of adjustment of the window setting. To obviate this problem, it is useful to obtain coronal sections; in coronal sections the difference between the signal intensity of the cerebellum and that of the normal cerebral hemispheres becomes quite obvious (Savoiardo et al., 1990) (Fig. 9B and D).

These atrophic changes and signal abnormalities correspond so exactly to the pathological cases of OPCA that we consider them pathognomonic. A clinical diagnosis of OPCA in a sporadic case, therefore, should not be accepted if it is not supported by the MRI findings we described, observed in more than 40 cases, almost all sporadic. Of course, there are borderline cases early in the course of the disease in which the clinical suspicion of OPCA does not correspond to definite MRI abnormalities; in a couple of such cases, however, repeat MRI 2 or 3 years later demonstrated a clear progression of atrophy and appearance of signal abnormalities, thus allowing the confirmation of the diagnosis (Fig. 8A and B).

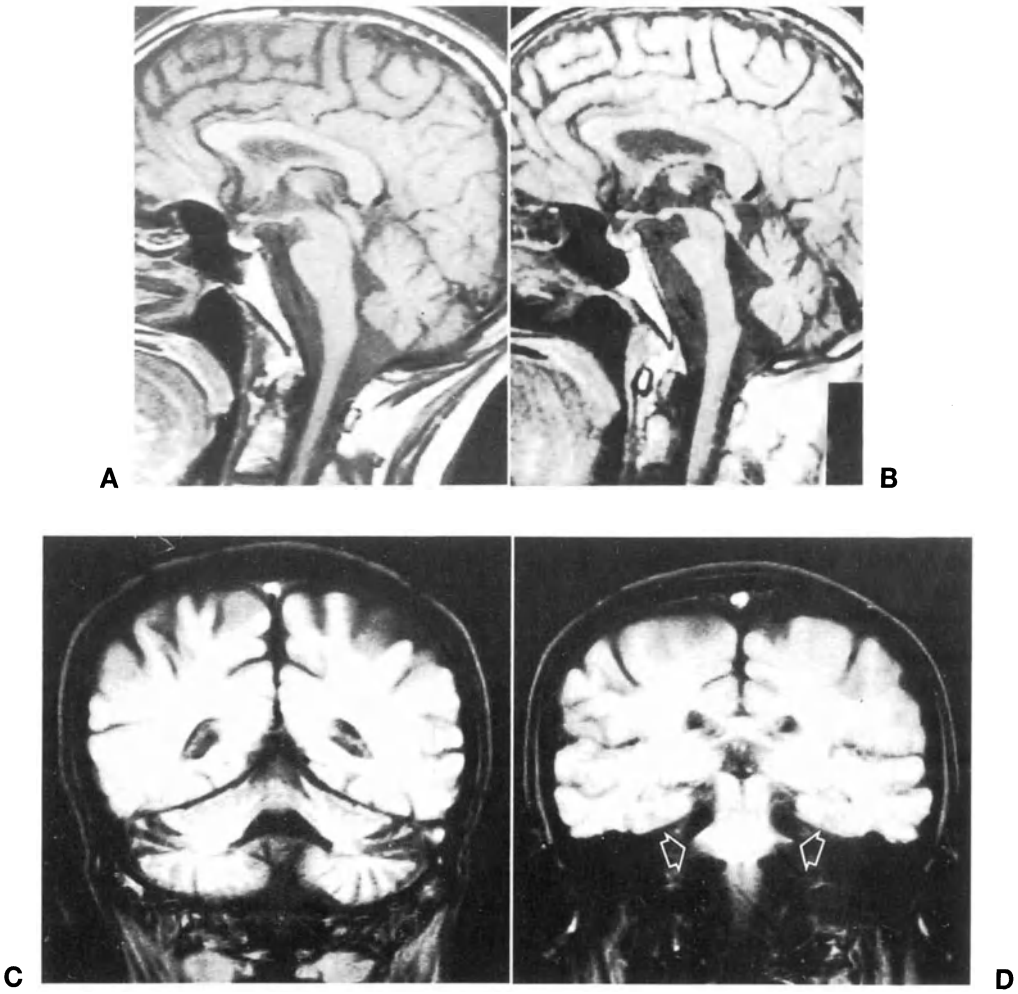

Fig. 8. Olivopontocerebellar atrophy. Early in the course of the disease, clinical and MRI findings may remain uncertain; note minimal flattening of the inferior part of the profile of the pons in **A**. Three years later, with clinically advanced disease, the atrophy of the pons and cerebellum is unquestionable (**B**) (both A and B, sagittal T1-weighted sections). T1-weighted coronal sections on the cerebellum (**C**) and pons (**D**) in a different patient demonstrate atrophy of the cerebellar hemispheres and of the middle cerebellar peduncles (open arrows)

MRI demonstration of multiple system atrophy

In a group of 11 MSA patients with marked cerebellar, autonomic, and extrapyramidal involvement, MRI abnormalities were present both in posterior fossa and in the putamen; there was MRI evidence, therefore, of involvement of multiple systems. However, in patients less severely involved, still classifiable as MSA, some discrepancies between clinical

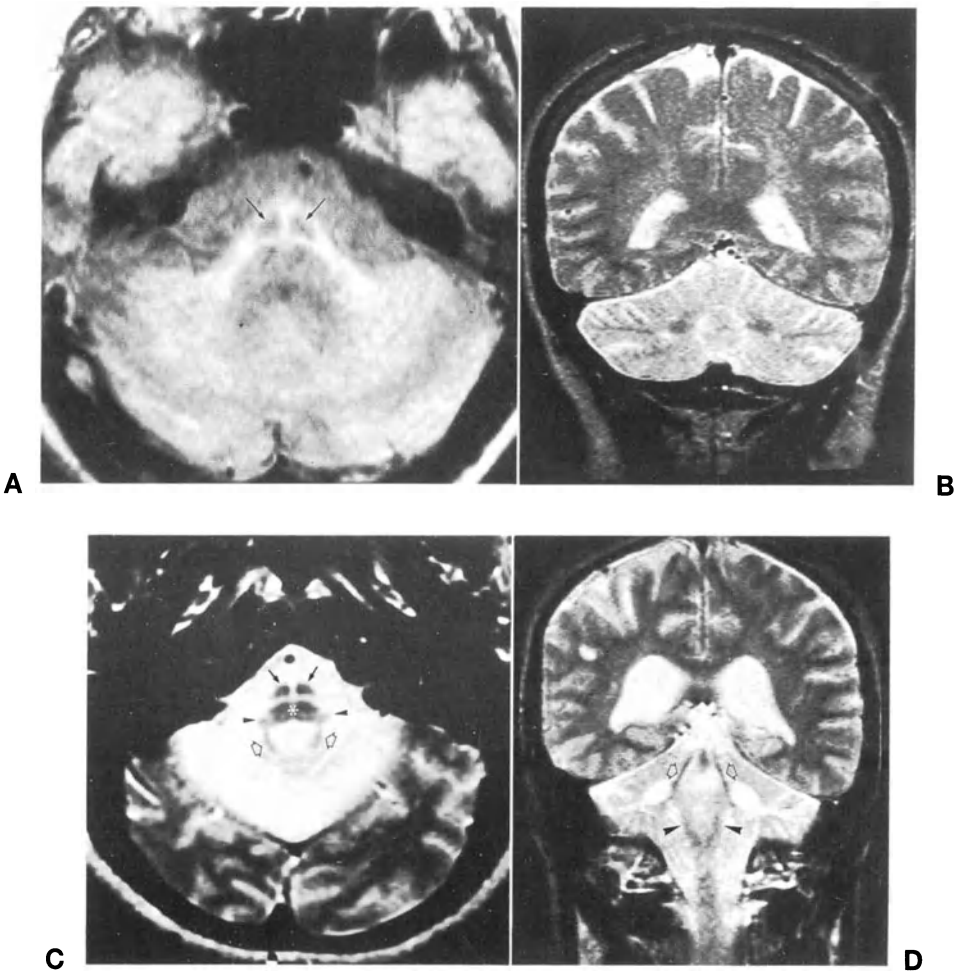

Fig. 9. Olivopontocerebellar atrophy. Signal hyperintensity is visible in T2-weighted and mainly in proton density images in the degenerate transverse pontine fibers and middle cerebellar peduncles (**A** axial proton density section; compare with Fig. 10; arrows indicate the normal pyramidal tracts). Cerebellar hyperintensity is particularly well demonstrated in coronal sections, by comparison with the cerebral hemispheres (**B**). The structures that in OPCA do not degenerate stand out by their normal signal intensity; these are: pyramidal tracts (arrows), pontine tegmentum (asterisk), inferior cerebellar peduncles (arrowheads), superior cerebellar peduncles (open arrows), (**C** T2-weighted axial section). The normal inferior (arrowheads) and superior (open arrows) cerebellar peduncles may be seen against the abnormal background in coronal section (**D**). [C and D, from Radiology (Savoiardo et al., 1990), with permission]

involvement of extrapyramidal or cerebellar systems and MRI demonstration of putaminal or posterior fossa involvement were observed.

In one case clinically labelled as OPCA, putaminal abnormalities were present on MRI, in addition to posterior fossa findings of OPCA; in 5 cases considered MSA but with not very severe extrapyramidal involvement,

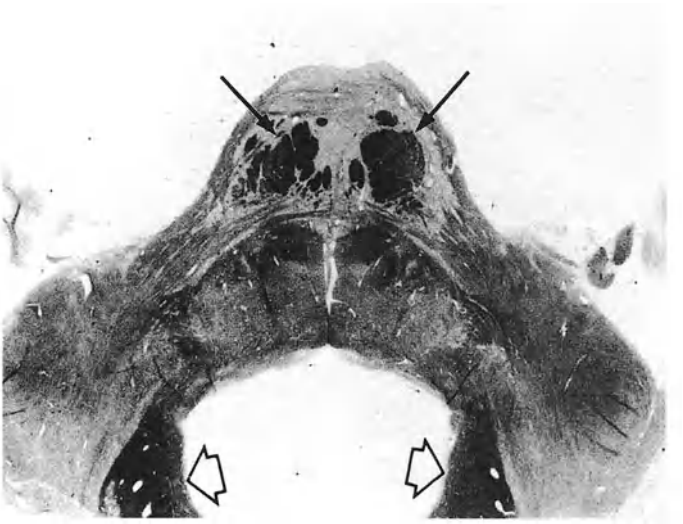

Fig. 10. Olivopontocerebellar atrophy. Axial section through the pons in a case of OPCA, stained for myelin (Woelcke-Heidenhain), demonstrates the validity of the MRI findings. Only the pyramidal tracts (arrows) and the superior cerebellar peduncles (open arrows) are normally stained, while the transverse pontine fibers are not stained. Compare with Fig. 9A and C. [Courtesy of Professor Orso Bugiani; from Radiology (Savoiardo et al., 1990), with permission]

MRI demonstrated cerebellar and pontine atrophy but no putaminal abnormalities. It is possible that MRI sensitivity is inferior to clinical sensitivity in the early diagnosis of extrapyramidal involvement.

Conclusions

In spite of the minor incongruities here described, MRI has become a very powerful tool in demonstrating abnormalities in posterior fossa in patients with OPCA. However, MRI sensitivity is inferior to that of clinical examination in the early diagnosis of several extrapyramidal disorders, but MRI can, indeed, provide evidence of putaminal involvement in the majority of MSA cases. It is also possible that other techniques such as PET studies may be more sensitive than MRI in the early detection of MSAs by demonstrating involvement of multiple systems in cases in which clinical and MRI evaluation indicates the involvement of only the cerebellar or the extrapyramidal system.

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