Short report

MRI in autonomic failure

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SUMMARY A significant rank correlation between rigidity and putaminal signal dropout on magnetic resonance imaging (MRI) in patients with multiple system atrophy suggests that putaminal degeneration may cause this clinical finding. Absence of putaminal abnormalities on MRI in patients with pure autonomic failure may prove useful in differentiating these two autonomic disorders.

Autonomic failure may occur alone (pure autonomic failure) or in association with central neurological disorders, including Parkinsonism, ataxia and pyramidal findings (multiple system atrophy).¹ Multiple system atrophy includes both striatonigral degeneration and olivopontocerebellar atrophy, since there is frequently overlap of patients in these two categories, and the clinical and pathological findings in multiple system atrophy encompass those described for both striatonigral degeneration and olivopontocerebellar atrophy.² The pathology of pure autonomic failure, however, has not yet been clearly defined.

Biochemical and pharmacological differences between multiple system atrophy and pure autonomic failure have increased our understanding of these diseases.¹ Unfortunately, these differences cannot be exploited as diagnostic tests in individual cases. The development of magnetic resonance imaging (MRI) has made it possible to examine structural details within the brain not visualised by computed tomography (CT) scans. We recently described putaminal atrophy on T₁ weighted axial scans of three of eight multiple system atrophy patients, and decreased signal intensity in the posterolateral putamen on T₂ weighted scans of seven of these patients by MRI,³ in

agreement with the cell loss seen pathologically in this area of the basal ganglia.² In this study, we compared MRI changes with clinical findings in these eight patients, and have used MRI to evaluate four patients with pure autonomic failure.

Materials and methods

Patients were referred to the National Institutes of Health (NIH) for evaluation of orthostatic hypotension. Medical history and neurological examination were performed to establish the diagnosis of pure autonomic failure or multiple system atrophy by criteria previously described.¹ The clinical characteristics of the multiple system atrophy patients have been reported previously.³ The pure autonomic failure patients ranged in age from 47 to 71 years, and had their illness for 9 to 12 years. No pure autonomic failure patient had signs or symptoms of nonautonomic neurological dysfunction. All subjects gave informed consent following a full explanation of the procedure in accordance with NIH guidelines.

Imaging was performed using a 1.5 T General Electric (Milwaukee, Wisconsin) Signa System. Images in the axial and coronal projections were obtained using a variety of T₁ and T₂ weighted spin-echo sequences (for example, TE 25/TR 600 ms and TE 70/TR 2000 ms respectively) with a nominal slice thickness of 0.5 cm. These were usually interleaved to provide maximum signal-to-noise ratio.⁴

Parkinsonism in the multiple system atrophy patients was ranked, using a modified Columbia scale (maximum score = 60),⁵ when patients were off medication for at least a week. Severity of putaminal changes on MRI was ranked independent of clinical data, and clinical and MRI scores were then compared by calculating Spearman's rank correlation coefficient.⁶

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Table Rank ordering of putaminal signal dropout and Parkinsonism in multiple system atrophy patients*

Patient No	MRI	Tremor	Rigidity	Bradykinesia	Overall Parkinsonism
1	1	1	1	1	1
2	8	4	7	2	4
3	4	6	4	4	5
4	2	3	2	5	3
5	5	5	6	6	6
6	6	2	3	3	2
7	7	8	8	7	8
8	3	7	5	8	7

*MRI scans are ranked in order of decreasing severity of putaminal signal dropout and Parkinsonism scores are ranked in order of decreasing clinical severity.

Results

Rank ordering of patients according to severity of MRI changes and Parkinsonism are given in the table. There was a significant rank correlation between clinical severity of rigidity in multiple system atrophy patients and degree of change in putaminal signal intensity ($r_s = 0.810$, $p < 0.05$). Correlations of tremor, bradykinesia and overall Parkinsonism with putaminal changes were not significant ($r_s = 0.357$, 0.048 and 0.381 respectively). MRI scans in the pure autonomic failure patients were normal.

Discussion

The clinical distinction between multiple system atrophy and pure autonomic failure may be impossible in the early stages, since multiple system atrophy patients may present solely with autonomic findings, and only later develop other manifestations of the disease. A correct diagnosis is important, however, because the prognosis of multiple system atrophy is markedly worse than that of pure autonomic failure. Brainstem auditory evoked potentials have been proposed as a means of early diagnostic separation of these two syndromes.⁷ If loss of signal intensity in the posterolateral putamen is unique for, or even more frequently encountered in multiple system atrophy, MRI appearance may also potentially serve to differentiate this syndrome from pure autonomic failure. It might be especially useful early in the course of the disease when other clinical manifestations may be minimal or lacking.

Lesions of the putamen in humans have been correlated with dystonia.⁸ In experimental rigidity produced in monkeys, lesions involved a number of brainstem structures, but not the putamen.⁹ The correlation between rigidity and putaminal changes in the present study, however, suggests a causative role

for putaminal degeneration in this aspect of Parkinsonism in multiple system atrophy.

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