Conventional and advanced MRI approach to movement disorders

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Learning objectives

The purpose of this paper is to:

• illustrate the wide magnetic resonance imaging (MRI) spectrum of rather subtle brain abnormalities in patients with various types of hyperkinetic or hypokinetic movement disorders,

• present potential pearls and pitfalls that can help to distinguish specific areas affected with neurodegeneration,

• discuss the advances in MRI technology that increase the sensitivity and specificity of the movement disorders detection.
Background

Primary (idiopathic) movement disorders are those with no detectable underlying cause like in idiopathic Parkinson's disease (IPD). Syndromes with findings other than pure movement disorder are describe as "plus syndromes" - progressive supranuclear palsy (PSP), multiple system atrophy (MSA) and corticobasal degeneration (CBD). Heredodegenerative disorders are those with a well-defined hereditary basis such as neurodegenerations with iron accumulations like in Huntington's disease (HD) and Wilson's disease. Secondary movement disorders are those with known identifiable causes such as tumors, stroke, injuries, vascular malformations, toxins, drugs and metabolic disorders.

The contribution of conventional MRI in the fields of vascular neurology, neuro-oncology, inflammatory and infectious disorders has been immeasurable during last several decades. However, the diagnostic value of this imaging modality was rather limited in detection of numerous neurodegenerative processes, including movement disorders. Typical imaging findings may help distinguish among disorders that have overlapping clinical presentations.
Findings and procedure details

This pictorial essay reviews the radiological MRI findings in patients scanned at our center, demonstrating the most commonly encountered neurodegenerative movement disorders. Structural and functional imaging features seen on MRI, magnetic resonance spectroscopy, diffusion-weighted imaging and iron imaging in patients with Parkinson disease and the Parkinsonian variant of multiple system atrophy, corticobasal degeneration and progressive supranuclear palsy are presented. Typical imaging characteristics of heredodegenerative disorders, such as Huntington's disease and Wilson's disease are illustrated. Secondary movement disorders, with known identifiable causes, such as tumors, stroke, injuries, vascular malformations, toxins, drugs and metabolic disorders are discussed.

Idiopathic Parkinson disease (PD)

Parkinson disease is a progressive neurodegenerative disorder caused by deficiency of the neurotransmitter dopamine as the consequence of degeneration of dopaminergic neurons in the substantia nigra (SN).

Clinically it is characterised by rigidity, tremor, bradykinesia and postural imbalance. The SN is a pair of tilted plate-like structures that lie within the midbrain. It is located between the crural fibers and the red nucleus. [1] The SN is divided into two parts: a dorsal part - pars compacta (SNc) and a ventral part - pars reticulata (SNr) (Fig. 1 on page 7).

In early stage of Parkinson disease, MRI findings does not indicate disease-specific changes so its main role is to rule out other underlying pathologies that could cause parkinsonisms. In advanced stage MRI reveals narrowing and eventually loss of the normal "swallow tail" appearance of the substantia nigra. [2] Pars compacta of the substantia nigra loses normal hyperintensity and their margins relative to ruber nucleus become blurred (Fig. 2 on page 7). There could be present cerebral atrophy but without pontine or midbrain atrophy. 1H MR spectroscopy reveals reduced putaminal tNAA levels in PD patients which could indicate mitochondrial dysfunction, comparing to healthy individuals. [3] On DTI studies, diffusivity of olfactory tracts is increased with decreased fractional anisotropy in SN and nigrostriatal tracts. [4]

Multiple system atrophy (MSA)

Multiple system atrophy is a degenerative neurological disorder affecting both autonomic nervous system and movement, caused by neurodegeneration in striatonigral or olivopontocerebellar structures. MSA is divided into two subtypes: MSA-P (MSA with predominant parkinsonism) and MSA-C (MSA with cerebellar ataxia). Specific MRI
findings in patients with MSA-P are putaminal atrophy with angulated margin of the lateral aspects of putamen instead of curvature margine. "Putaminal rim sign" represents thin rim of hyperintensity of the posterolateral aspect on T2-weighted images due to gliosis (Fig. 3 on page 8, Fig. 4 on page 9). Medial to this posterolateral hyperintensity is a low intensity region on T2W and SWI images due to iron deposition (Fig. 5 on page 10). Image findings could be unilateral at the early stages of the disease and usually starts at the posterolateral region and later, signal changes extends to the anterior parts of the putamen. [5,6] SWI can visualize putaminal atrophy and marked signal hypointensity in patients with parkinsonism-predominant multiple system atrophy with high specificity.

The pathognomonic changes in MSA-C are degeneration of the pontocerebellar tract and pontine nuclei. Typical findings shows atrophy in brainstem with a cruciform pontine hyperintensity due to gliosis of transverse fibres and median raphe knows as the "hot cross bun sign" (Fig. 6 on page 11).

**Progressive supranuclear palsy (PSP)**

Progressive supranuclear palsy is characterized by slowness, rigidity, bradykinesia, repeated falls, downgaze limitation and dementia. Midbrain atrophy with sparing of pons on MRI is highly suggestive of PSP and is described as "hummingbird sign" (Fig. 7 on page 12, Fig. 8 on page 13). This sign is very helpful in differentiating PSP patients from those with Parkinson's disease and multisystem atrophy. [7] Another significant sign is "Mickey Mouse appearance" in which exists reduction of anteroposterior midline midbrain diameter, at the level of the superior colliculi on axial imaging (Fig. 9 on page 14).

**Corticobasal degeneration (CBD)**

Corticobasal degeneration is a progressive neurodegenerative disorder with extrapyramidal, cognitive and neuropsychiatric dysfunctional symphomatology, caused by asymmetric neuronal loss most severely in the posterior frontal and parietal lobes with degeneration of the corticospinal tract and basal ganglia. MRI may show asymmetrical cortical atrophy, T2W and PD hyperintensities in affected cortical grey matter and underlying white matter, atrophy of the central part of the corpus callosum, T2W hyperintensity of the putamen and globus pallidus and diffuse supratentorial atrophy without midbrain atrophy. In the absence of the structural MRI changes, a helpful tool is using DTI tractography which should show asymmetric increase in mean diffusivity in posterior frontal and anterior temporal regions. Volumetric MRI is more superior in finding regional differences in volume loss which may help identify CBD earlier. [8]

**Other causes of parkinsonism (secondary parkinsonism):**
• Infectious - abscess (Fig. 10 on page 15), encephalitis lethargica and other viral infections (e.g. AIDS, progressive multifocal leukoencephalopathy), prion disease, neurosyphilis, toxoplasmosis

• Toxic - carbon monoxide (Fig. 11 on page 16), cyanide, carbon disulfide, manganese

• Drug - induced dopamine receptor blockers, classic neuroleptics, atypical antipsychotics, other drugs

• Brain tumours: supratentorial and brainstem tumours (Fig. 12 on page 17), arteriovenous malformations

• Cranial trauma - chronic subdural haematoma, midbrain trauma, vascular lesions

• Metabolic - hypoxia, hypoparathyroidism, familial basal ganglia calcification, extrapontine myelinolysis, chronic liver failure, Wilson's disease (Fig. 13 on page 17, Fig. 14 on page 18)

• Miscellaneous - Huntington's disease (Fig. 15 on page 19), SCA mutations, FTDP-17, neuroacanthocytosis, normal-pressure hydrocephalus. [9]
Fig. 1: Axial T2W MR image showing normal anatomy of the substantia nigra through the upper midbrain. The hypointense areas represent the SNr and the red nucleus because of their high iron concentration. A band of relative hyperintensity between the hypointense SNr and red nucleus represents the SNc, which contains half the iron of the SNr.

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**Fig. 2:** Axial T2W MR image reveals bilateral narrowing of normal hyperintens signal of the pars compacta with blurred margins relative to nucleus ruber.

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**Fig. 3:** Axial T2W MR image shows bilateral putaminal atrophy with low signal intensity of the posterolateral segments of putamen with surrounding hyperintense rim.

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**Fig. 4:** Axial T2W MR image shows bilateral putaminal atrophy with low signal intensity of the posterolateral segments of putamen.

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Fig. 5: Axial SWI MR image shows bilateral putaminal atrophy with significant signal hypointensity of posterolateral aspects of the putamen.

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Fig. 6: Axial T2W MR image shows atrophy in brainstem that contains vertical and horizontal lines representing gliosis in the transverse fibers and median raphe compatible with the 'hot cross bun' sign.

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Fig. 7: Midsagittal T1W MR image shows normal size of the midbrain at the level of the interpeduncular cistern.

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**Fig. 8:** Midsagittal T1W MR image shows atrophy of the midbrain at the level of the interpeduncular cistern with the characteristic 'hummingbird' appearance.

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**Fig. 9:** Axial T2W MR image shows reduction of anteroposterior midline midbrain diameter, at the level of the superior colliculi on axial imaging (diameter below 12mm) giving the characteristic 'morning glory' or 'Mickey Mouse' appearance.

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Fig. 10: Axial T2W and contrast-enhanced axial T1W shows right frontal abscess.

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**Fig. 11:** Axial T2W and coronal T2W MR images shows bilateral high signal intensity in the affected globus pallidus due to carbon monoxide poisoning.

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**Fig. 12:** Axial T2W MR images shows thalamic tumor and a supratentorial brain tumor in a different patients.

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Fig. 13: Axial T2W MR image shows pathological high signal in the basal ganglia (putamen, globus pallidus and caudate nucleus) and anterolateral aspect of the thalamus, giving the characteristic ‘panda sign’ appearance.

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Fig. 14: Axial SWI MR images shows paramagnetic deposition in the globus pallidus and midbrain nuclei, related to cooper or iron accumulation in a patient with Wilson disease.

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Fig. 15: Contrast-enhanced T1W coronal MR image (A) and axial T2W FLAIR image shows caudate nucleus head and putaminal atrophy resulting in enlargement of the frontal horns.
Conclusion

Detection of subtle signs on conventional MRI and introduction of the "next level" imaging technology not only increase the diagnosis of movement disorders, but also improve our understanding of the underlying mechanisms.
References


